# Pediatric Surgery

## Diagnosis and Treatment

Christopher P. Coppola
Alfred P. Kennedy Jr
Marc S. Lessin
Ronald J. Scorpio
*Editors*

*Second Edition*

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# Foreword

It's 3 AM on a Saturday and, of course, it's February with ample snow on the ground. The 9-month-old you admitted with dehydration and vomiting is a former 28-week infant who was thriving…until tonight. You examine the abdomen once more, re-look at the X-ray, see the rise in the lactate, and now know you have crossed over the medical chasm—you need a pediatric surgeon.

As a pediatric critical care physician, I am fortunate to know unique individuals who have mastered the discipline of pediatric surgery—those physicians who can easily engage and comfort a distressed child (and family) *and* can adeptly diagnose, repair, and/or remove the source of a child's distress.

The editors are all close friends and colleagues. They are kind, patient-centered, and highly accomplished surgeons with over 75 years of cumulative experience. I have been elbow-to-elbow with each of these surgeons on many a February night.

However, I wondered why four of the busiest people I know would take on a textbook. Then I was struck by a line in the preface of the first edition "*Every time a pediatrician diagnoses a child with abdominal pain, an emergency medicine physician cleans a child's wound, or a nurse chooses a location for an intravenous line in an infant, pediatric surgical care is being delivered*".

I believe this summarizes the intent of the editors—a desire to educate providers across skill sets so to optimize the care of any infant or child with a potential surgical disorder. This is not surprising and reflects the generosity, lack of hubris and the expertise these thoughtful editors demonstrate day in and day out…nights included.

What a blessing to know a good pediatric surgeon.

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# Preface

The surgical care of children is often a source of worry for medical team members due to the high stakes involved. Injury and surgical illness can result in critical conditions in children requiring rapid assessment and action to achieve a return to health. It is important to remember when there is a sick child, there is a sick family, and the needs of parents must be addressed. Pediatric surgery as a field is challenging due to its breadth. It remains a generalized discipline in which the conditions treated span literally head to toe. It is oft repeated that children are not small adults. The surgical conditions of children are distinct from adults, and signs and symptoms in children indicate different etiologies. A relatively small number of pediatric surgeons are concentrated in tertiary treatment centers so children with surgical disease will often present to non-pediatric specific providers.

This text is designed to assist those caring for children whether they are on a pediatric surgery team or not. It is organized by specific surgical diseases affecting children at each stage of life, as well as chapters on trauma, neoplasm, and technical procedures. Chapters organize information in outline form to enable rapid diagnosis and treatment. Chapter structure includes key points, a narrative section of additional details, study questions, and further reading. Each topic is written to stand alone for rapid reference, or the text can be used as a whole for study. We editors have devoted our professional lives to improving the lives of children and hope that this work helps others help children in need.

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# Part I
# Introduction

# The History of Pediatric Surgery

**Max D. Hazeltine and Michael P. Hirsh**

## Origins

The care of children in a specialized institution dates back to at least 1739, when Thomas Coram established the Foundling Hospital in London. By 1769 George Armstrong organized a clinic in London dedicated to caring for children's diseases, called the Dispensary for the Infant Poor. Over the ensuing century, a number of clinics and hospitals devoted to children were founded internationally, with L'Hôpital des Enfants Malades opening in Paris in 1802, the Hospital for Sick Children in London in 1852, a 12-bed Children's Hospital in Philadelphia in 1855, and a 20-bed Children's Hospital in Boston in 1869. Further developments were seen in 1860, as the first textbook on children's surgery was published by J. Cooper Forster. Additionally, the advent of anesthesia in 1846 enabled surgeons to perform more complex surgical procedures, further accelerating the development of the field of surgery.

Despite these advances in the care of children in the nineteenth century, there was a schism in the philosophy of how to care for medical versus surgical diseases. Dr. Charles West, who was a founder of the Hospital for Sick Children in London, believed that children with diseases that were medical in nature required specialized attention, while surgical disease should be treated in general hospitals. The Hospital for Sick Children in London followed this philosophy, and a number of British hospitals followed suit. However, in the latter part of the nineteenth century, surgery in

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children became more recognized as requiring specialized facilities and care, particularly after the invention of anesthesia. This was demonstrated after the Royal Edinburg Hospital for Sick Children created a ward for surgical patients in 1887 and converted a sewing room into an operating theater. In fact, Charles Dickens was a major supporter of creating a facility for all children who needed specialized care. This progressed to specialized training in pediatric surgery that was independent of general surgery. By the 1890s St. Mungo's College in Glasgow was providing pediatric surgery lectures ranging from fractures to congenital malformations.

## Development into a Specialty

Pediatric surgery began to take shape during the early 1900s. One of the most prominent figures was William E. Ladd, born in 1880, who was practicing in Boston. His practice involved both children and adults, but this changed drastically after the famous Halifax disaster on December 6, 1917, in which he led a medical mission to treat children injured by the explosion. One must note, however, that Dr. Ladd had already been making significant strides as a visiting staff to Boston Children's Hospital since 1910. He and Dr. James Stone halved the mortality rate from intussusception by utilizing early diagnosis and surgical intervention.

While Dr. Ladd is widely recognized as the godfather of pediatric surgery, he was not the first surgeon to devote his entire practice to pediatric surgery. That recognition goes to Dr. Herbert Coe of Seattle. After World War I, Dr. Coe visited Boston Children's Hospital and trained alongside Dr. Ladd. Coe then returned to Seattle and by 1919 was exclusively practicing pediatric surgery at the Children's Orthopedic Hospital.

By 1927 Ladd devoted his entire practice to children despite the financial disadvantage from closing his adult practice. His growing expertise began to demonstrably improve the care of children, with his publications documenting improvements in the care of a variety of diseases ranging from intussusception, pyloric stenosis, duodenal obstruction and malrotation, to biliary atresia. Similar advancements were made internationally, with Gertrude Herzfeld from the Royal Hospital for Sick Children in Edinburgh reporting her experience of 2100 hernia repairs over 5 years. She advocated for repair at any age and even performed surgery within patient's homes.

After Ladd became fully devoted to pediatric surgery, he began to recruit surgeons from other specialties to carve out similar niches. He first recruited Thomas Lanman in urology in 1927, Franc Ingraham in neurosurgery in 1929, Carlyle Flake in otolaryngology in 1937, and Donald MacCollum in plastic surgery in the late 1930s. During the 1930s Robert Gross emerged into the world of pediatric surgery. He sought a surgical internship, and a 67-word letter of recommendation from Charles McKhann to Dr. Ladd noted that Gross was "…somewhat above the average…," and "…has a pleasant personality and a good appearance. He should make a satisfactory house officer." Gross did not obtain the surgical internship and went on to train in pathology. However, he eventually made his way into the field of surgery where he trained at Brigham Hospital as well as Children's Hospital under Ladd. It was during his time at Boston Children's Hospital that Gross performed the first elective cardiovascular operation in history in 1938, the ligation of a ductus arteriosus in a 7-year-old girl. He performed the operation in defiance of Ladd's command to not do the procedure while he was away on vacation. This led to Ladd attempting to fire Gross, which strained the relationship so significantly that some felt it hindered the advance of the field of pediatric surgery. Nevertheless, Gross went on to succeed Ladd as the surgeon-in-chief after Ladd's retirement.

The foundations laid in the preceding decades led to significant advances in the field of pediatric surgery in the 1940s. In 1941 Ladd and Gross published the first modern pediatric surgery textbook, *Abdominal Surgery of Infants and Children*, which, among other major operations, described the Ladd operation for malrotation of the intestine. Cameron Haight performed the first successful correction of esophageal atresia with distal tracheoesophageal fistula in 1941. The first bypass operation for tetralogy of Fallot was performed by Alfred Blalock, Vivien Thomas, and Helen Taussig in 1944. The first effective operation for Hirschsprung disease was performed in 1948 by Orvar Swenson, and Mark Ravitch described the barium enema reduction of intussusception, mucosal proctectomy and ileoanal pull-through for benign colon disease, as well as the pectus excavatum repair. A number of major children's hospitals were also being formed, such as Children's Hospital of Philadelphia, Children's Memorial Hospital in Chicago, as well as programs in Toronto and Montreal. A number of these hospitals began offering formalized training experiences in pediatric surgery.

Perhaps the most influential of accomplishments during this time was that of Dr. Coe, who organized the first professional society of pediatric surgeons in 1948, the Section on Surgery under the American Academy of Pediatrics. Internationally, the field saw the first meeting of the Scottish Surgical Paediatric Club in 1948, as well as the formation of the British Association of Paediatric Surgeons in 1953. Despite these achievements, C. Everett Koop, a prominent pediatric surgeon at the Children's Hospital of Philadelphia, faced significant resistance in establishing a formal certifying board of pediatric surgery. His application before the Advisory Board for Medical Specialties (ABMS) was rejected in 1957, 1961, and 1967. After the 1961 rejection, the ABMS remarked to Koop that "You have no body of knowledge, no certified training programs, and no journal." The field would promptly reply. By 1965 William Clatworthy's efforts led to the Clatworthy report, outlining the standards for basic principles and requirements for the education of pediatric surgeons, which 11 programs in the United States and 2 in Canada met [1, 2]. The *Journal of Pediatric Surgery* ran its first issue in February 1966, with C. Everett Koop as its Editor-in-Chief. Koop would later go on to become Surgeon General of the United States. The Canadian Association of Paediatric Surgeons was formed in 1967. However, the ABMS again rejected certification status in pediatric surgery in 1967, and the American Board of Surgery (ABS) chair felt that a strong national and independent pediatric surgery organization would be essential to the future of pediatric surgery. In response, the American Pediatric Surgical Association (APSA) was formed in 1970 with 191 members and Dr. Gross its first president. Finally, in 1973 the ABS granted a new certificate of special competence in pediatric surgery to be awarded to qualified applicants.

The first official board examination in pediatric surgery would occur in Puerto Rico in 1975, just prior to the sixth APSA meeting. The goal was to certify the approximately 300 surgeons in active practice. Ultimately 250 surgeons took the 3-hour exam, with 238 passing. The only two individuals to not take the exam were Gross and Swenson, who were grandfathered in. They received the first and second diplomas, respectively, with Dr. Judson Randolph having said, “We did not have the temerity to examine those two.”

## Refinement and Growth of Pediatric Surgery into the Modern Era

After its recognition by the ABS, the next major step was formalizing and expanding training programs. The Clatworthy committee that was initially created in the mid-1960s to evaluate training programs continued to do so, and these programs were then approved by the ABS. By the late 1970s, this evaluation process was taken over by the Liaison Committee on Graduate Medical Education and the Residency Review Committee for Surgery. In 1978 there were ten approved training programs. The precursor to the Association of Pediatric Surgery Training Program Directors was created in 1976 and incorporated in 1989.

Although officially recognized, there were still challenges met by the generation of pediatrics surgeons who received the first formal training. Pediatricians were unsure of how pediatric surgeons would benefit their patients. General surgeons were offended by the idea that pediatric surgeons could do a better job caring for patients than them. Judson Randolph, after arriving at the Children’s Hospital of the District of Columbia, was met with attempts to oust him after a baby died from complications after his esophageal atresia repair. Adult surgeons requested the hospital formally investigate the case for negligence and incompetence. Their attempts were unsuccessful after Dr. Randolph pointed out that none of these surgeons had a patient survive after performing their own repairs. Randolph went on to become president of APSA from 1984 to 1985.

Nevertheless, the field marched on. The advances in pediatric surgery since the 1960s were innumerable. Below are a variety of achievements that have led to the success of the field as a whole.

- *1968*: Morio Kasai provided his first English report of hepatic portoenterostomy for biliary atresia in the *Journal of Pediatric Surgery*. This surgery led to jaundice-free survival in 55% by the mid-1980s, which was previously a fatal disease.
- *1968*: Douglas Wilmore and Stanley Dudrick reported on the first patient sustained by total parenteral nutrition, a newborn infant with near-total atresia of the small bowel.

- *1968*: Purushottam Upadhyaya and Jones Simpson of the Hospital for Sick Children in Toronto described the selective management of splenic injuries. This eventually led to Richard Pearl's 1998 update that reported an overall salvage rate of 96%, leading to a paradigm shift for both pediatric and adult trauma patients.
- *1969*: Helen Noblett, who practiced in Australia and England, developed the use of water-soluble radiological contrast for meconium ileus and the use of a suction rectal biopsy to diagnose Hirschsprung disease.
- *1969*: The first African American pediatric surgeon, James Densler, started practice in Atlanta. He was the second African American to receive board certification in pediatric surgery by the ABS but was never an APSA member until receiving an honorary membership in 2020.
- *1971*: Stephen Gans described the use of "peritoneoscopy," later known as laparoscopy, in infants and children.
- *1974*: Barbara Barlow published how breast milk prevented rat infants from developing necrotizing enterocolitis.
- *1977*: Lester Martin described his approach to mucosal proctectomy, total colectomy, and ileoanal pouch anastomosis for ulcerative colitis, modernizing the treatment strategy of ulcerative colitis.
- *1981*: The first open fetal surgery is performed by Michael Harrison.
- *1982*: Alberto Peña presented his novel surgical approach to imperforate anus to APSA.
- *1991*: The first reports of a laparoscopic cholecystectomy performed in children are disseminated.
- *1992*: First reports of therapeutic fetoscopy are reported, although diagnostic procedures had been performed as early as the 1960s.
- *1994*: The laparoscopic Swenson abdominoperineal pull-through is performed in dogs to assess its feasibility for use in children.
- *1995*: The first reports of a robot-assisted thoracoscopic surgery are published.
- *2000*: The first pediatric cases using a fully integrated robotic platform were performed.
- *2003*: Heung Bae Kim described the serial transverse enteroplasty, a novel bowel lengthening procedure.

One can easily see that pediatric surgery, while in its infancy as a field, had a meteoric rise to prominence. From its beginnings as a part-time practice for a handful of surgeons, by 2019 there were 1481 members in APSA and 59 accredited training programs.

## The Future of Pediatric Surgery

Pediatric surgery is now one of the most sought-after specialties. The field attracts top candidates and has become one of the most competitive specialties to enter. However, concerns exist regarding the appropriate balance for the workforce. Some evidence exists that there are more surgeons trained than there are children with rare index surgical cases being born, which has led to dilution of cases among existing surgeons and trainees. Advances in medical treatments have also prevented the formation of surgical diseases such as spina bifida, as well as have become so effective that surgical intervention has become less frequent. Data have shown that more mundane cases (e.g., appendectomy, umbilical hernia repair) and fewer index cases (e.g., diaphragmatic hernia repair, pectus deformity repair) are being performed, which some fear may lead to a lack of proficiency.

The way care is delivered has also changed with the evolution of modern healthcare. Care may be migrating to high-volume, specialty-specific centers. Complex cases may be seen at large public hospitals, while elective cases are more often performed in private institutions without trainee coverage. Furthermore, workforce needs vary by region and country. Rural areas and developing nations need more pediatric surgeons, while some major urban centers have an excess of surgeons.

Surgical innovation has continued forward with more procedures being performed by a minimally invasive approach. Laparoscopic surgery has revolutionized the field of pediatric surgery, adding new approaches to the pediatric surgeon's armamentarium and allowing faster recovery for patients. Robotic surgery is another blossoming technology that is rapidly advancing the field of pediatric surgery. Within the span of a decade, the number of published robotic surgical procedures in children has gone from nearly zero to over 1300. As innovation continues to grow, we will see significant changes in surgical approach and the resultant benefits for children.

In addition to surgical innovation, novel collaborations have enhanced care for children in the United States and worldwide. The Children's Oncology Group (COG), formed in 2000, is the world's largest research organization dedicated exclusively to children and adolescents with cancer. COG includes over 200 hospitals, universities, and cancer centers across the world. Their research efforts in children's cancer have helped improve survival from almost incurable decades ago to nearly 80% today.

The field of pediatric surgery is ripe with surgeon-scientists, with proportionally more than other surgical specialties. Pediatric surgeons, as compared to other surgical specialties, more often felt surgeons can succeed in today's challenging research environment and reported the highest rate of National Institutes of Health funding. The pediatric surgeon-scientists have pioneered research in areas such as regenerative medicine, tissue engineering, the enteric nervous system, necrotizing enterocolitis, and wound healing. Additionally, gene therapy has been emerging as a novel therapy for a variety of diseases. The first Food and Drug Administration (FDA)-approved gene therapy came in 2017, with Kymriah, a genetically modified T-cell for the treatment of B-cell acute lymphoblastic leukemia. Currently, there are 20 FDA-approved cellular and gene therapy products.

Whatever may lie in store for the field, it will be met with the same enthusiasm and tenacity that helped found it. From its inception, where it was viewed as a financially unviable field and a secondary part of a surgeon's practice, pediatric surgery has overcome a myriad of obstacles to become one of the preeminent medical specialties. We can learn from the contentious relationship between Ladd and Gross, which Koop felt significantly obstructed the field's progress. Rather, the emphasis should be on unity for the future of children's health.

## References

1. Grosfeld JL, O'Neill JA, Coran A, Fonkalsrud EW. Pediatric surgery: Sixth edition. Sixth ed. Philadelphia, PA: Mosby Elsevier; 2006. p. 1–2368.
2. Nakayama DK, Powell D, Fallat ME, Holcomb GW 3rd. Saving lifetimes: celebrating the 50th Anniversary of the American Pediatric Surgical Association, 2019. 149p.

## *Further Reading*

Administration USFaD. FDA approval brings first gene therapy to the United States. 2017.

Administration USFaD. Approved cellular and gene therapy products, 2021. 27 March 2021.

Beasley SW. The challenges facing training in pediatric surgery worldwide. Front Pediatr. 2013;1:24.

Bill AH, Herbert E. Coe, 1881–1968. J Pediatr Surg. 1969;4(1):1–2.

Bruns NE, Shah MA, Dorsey AN, Ponsky TA, Soldes OS. Pediatric surgery—a changing field: national trends in pediatric surgical practice. J Pediatr Surg. 2016;51(6):1034–8.

Bunim J. UCSF surgeon reflects on performing world's first fetal surgery 30 years ago 2011–12. May 2021. https://www.ucsf.edu/news/2011/02/103676/ucsf-surgeon-reflects-performing-worlds-first-fetal-surgery-30-years-ago

Codsi E, Audibert F. Fetal surgery: past, present, and future perspectives. J Obstet Gynaecol Can. 2019;41:S287–S9.

Crombleholme TM, Johnson MP. Fetoscopic surgery. Clin Obstet Gynecol. 2003;46(1):76–91. https://doi.org/10.1097/00003081-200303000-00011.

Cundy TP, Shetty K, Clark J, Chang TP, Sriskandarajah K, Gattas NE, et al. The first decade of robotic surgery in children. J Pediatr Surg. 2013;48(4):858–65.

Curran TJ, Raffensperger JG. The feasibility of laparoscopic swenson pull-through. J Pediatr Surg. 1994;29(9):1273–5.

Estes JM, MacGillivray TE, Hedrick MH, Adzick NS, Harrison MR. Fetoscopic surgery for the treatment of congenital anomalies. J Pediatr Surg. 1992;27(8):950–4. https://doi.org/10.1016/0022-3468(92)90538-i.

Fallat ME. Redefining Ladd's path. J Pediatr Surg. 2017;52(1):3–15.

Group CsO. About us 2021. https://www.childrensoncologygroup.org/index.php/about-us

Group TCC. About us 2021. https://www.coram.org.uk/about-us

Hendren WH, Folkman MJ. A Memorial Minute: Robert Edward Gross. https://fa.hms.harvard.edu/files/memorialminute_gross_robert_e.pdf

Holcomb GW 3rd. The Journal of Pediatric Surgery: its first 50 years. J Pediatr Surg. 2018;53(2):212–6.

Hospital BCs. Boston Children's Hospital History. 2021. https://bcrp.childrenshospital.org/history/boston-childrens-hospital/

Kim HB, Lee PW, Garza J, Duggan C, Fauza D, Jaksic T. Serial transverse enteroplasty for short bowel syndrome: a case report. J Pediatr Surg. 2003;38(6):881–5.

Markel H. Charles Dickens' work to help establish Great Ormond Street Hospital, London. Lancet. 1999;354(9179):673–5.

Meininger D, Byhahn C, Markus BH, Heller K, Westphal K. Total endoscopic Nissen fundoplication with the robotic device “da Vinci” in children. Hemodynamics, gas exchange, and anesthetic management. Anaesthesist. 2001;50(4):271–5.

Nakayama DK. Vignettes from the history of pediatric surgery. J Pediatr Surg. 2020;55s:1–37.

Okada S, Tanaba Y, Yamauchi H, Sato S. Single-surgeon thoracoscopic surgery with a voice-controlled robot. Lancet. 1998;351(9111):1249.

Philadelphia CsHo. About the History of Children’s Hospital. 2021. https://www.chop.edu/about-us/about

Robinson DH, Toledo AH. Historical development of modern anesthesia. J Investig Surg. 2012;25(3):141–9.

Sigman HH, Laberge JM, Croitoru D, Hong A, Sigman K, Nguyen LT, et al. Laparoscopic cholecystectomy: a treatment option for gallbladder disease in children. J Pediatr Surg. 1991;26(10):1181–3.

Steen EH, Moles CM, Goldstein AM, Morowitz MJ, Gosain A, Mollen KP, et al. The pediatric surgeon-scientist: succeeding in today’s academic environment. J Surg Res. 2019;244:502–8.

Withycombe JS, Alonzo TA, Wilkins-Sanchez MA, Hetherington M, Adamson PC, Landier W. The children’s oncology group: organizational structure, membership, and institutional characteristics. J Pediatr Oncol Nurs. 2019;36(1):24–34.

Zeller KA, Nakayama DK, Fraile K, Held JM, Simpson JL, Koehler S, et al. History of pediatric surgery. pediatric surgery nat, american pediatric surgery association and unbound medicine. 2020. https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829734/all/HistoryofPediatricSurgery. Accessed 2 May 2022.

# Resources for Pediatric Surgery

## Kelly Smith

**Key Points**

1. National Surgical Quality Improvement Program, Pediatric (NSQIP-P) is a national quality data program through the American College of Surgeons which helps participating centers compare their surgical outcomes data with each other in a risk-adjusted manner. It gives the participating center reliable data from which to determine opportunities for improvement of surgical care for children.
2. The Children's Surgery Verification Program is a national program designated by the American College of Surgeons where facilities can apply to become verified as a Level I, II, or III center.
3. High demand for pediatric surgeons which exceeds supply in many communities creates a need for either transfer out of children with complex conditions or employment of locum tenens pediatric surgeons to make continuous pediatric surgical services available.

1. NSQIP pediatric
    a. The American College of Surgeon's National Surgical Quality Improvement Program (ACS NSQIP) allows participating centers to "collect highly reliable clinical data and compare their surgical outcomes with the outcomes of other participants in the program." This program is the "first national data-

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base to measure surgical outcomes in all specialties of children's surgical care" (Surgeons, American College of Surgeons, 1996–2021).

   b. NSQIP Pediatric (NSQIP-P) requires a full-time Surgical Clinical Reviewer who reviews each chart based on inclusion criteria of the program for around 230 data points of preoperative, intraoperative, and postoperative variables.
   c. The data collection includes 15 surgical specialties from which cases are selected:

      i. Pediatric Cardiovascular-Thoracic Surgery
      ii. Pediatric Neurosurgery
      iii. Pediatric Orthopedic Surgery
      iv. Pediatric Otolaryngology
      v. Pediatric Surgery
      vi. Pediatric Urology
      vii. Pediatric Plastic Surgery
      viii. Plastic Surgery
      ix. Cardiovascular-Thoracic Surgery
      x. General Surgery
      xi. Gynecology
      xii. Neurosurgery
      xiii. Orthopedic Surgery
      xiv. Otolaryngology
      xv. Urology

   d. Participating centers receive a risk-adjusted report semi-annually to compare their outcomes against other participating pediatric surgical centers.
   e. Report data can be utilized by centers to develop quality improvement projects based on their outlier status as compared to other participating centers with many occurrences such as surgical site infection (SSI), venous thromboembolism (VTE), pneumonia, sepsis, etc. These reports show participating centers if they are in the average, above average, or below average group for tracked complications.

2. Children's surgery verification

   a. "Children's Surgery Verification (CSV) through the American College of Surgeons (ACS) is the nation's first and only multispecialty standards for children's surgical care" [1]. This program was developed from a task force for Children's Surgical Care and was composed of a multidisciplinary group of leaders in children's surgical care and participation and support of the ACS [2]. The mission of the verification program is to "improve the quality of children's surgical care by creating a system that allows for a prospective match of every child's individual surgical needs with a care environment that has optimal pediatric resources" [1].

b. Application to the Children's Surgery Verification program is available to any center that cares for children and includes specialty hospital verifications for Musculoskeletal and Oncology centers.
c. There are three levels of verification award that can be obtained by meeting the standards set forth by the American College of Surgeons to demonstrate optimal quality of care of children's surgical patients.
d. Level 1 verification is the highest level of verification and includes patients of any age. Level 1 centers can care for the most complex patients with ASA 1–5. Multiple medical and surgical specialties and pediatric anesthesiology are required for multidisciplinary patient management. Operations can include procedures for major congenital anomalies and complex diseases, including those that are uncommon or require significant multidisciplinary coordination [3].
e. Level 2 verification includes patients of any age. Level 2 centers can care for patients with ASA 1–3 and are typically single surgical specialties and have neonatology and pediatric anesthesiology providers available. Operations can include procedures for common anomalies and diseases that are typically treated by most children's surgical specialists and that do not require significant multispecialty coordination [3].
f. Level 3 verification includes patients greater than 6 months of age with an ASA of 1–2. There is no multidisciplinary management of comorbidities required. Operations include common, low-risk procedures typically performed by a single specialty [3].
g. Prior to application for Children's Surgery Verification, a center must ensure they meet the requirements and standards of the program for the level that they are applying, as delineated in the Optimal Resources for Children's Surgical Care. Creating a gap analysis is a helpful tool to analyze the standard requirements and determine any areas that may need focused on. Our center utilized a red/yellow/green color code system to signify areas on the gap analysis to identify areas for improvement or growth to meet the required standards.
h. Centers applying for Level 1 or Level 2 verification are required to participate in NSQIP-P for 1 year prior to application. Also, programs must have quality improvement projects derived from their surgical quality data to show at the time of site visit. Data collection for NSQIP-P requires the center to employ a full-time Surgical Clinical Reviewer to review all included surgical charts and enter them into the NSQIP-P database for comparison to other centers nationally on the Semi-Annual Report (SAR) that is released every January and July each year.
i. A multidisciplinary group of surgeons is required for comprehensive surgical care of children. Children's Surgery Verification requires at least one pediatric general surgeon on staff for Level 2 verification. Level 1 verification requires two or more pediatric general surgeons.

j. A Performance Improvement and Patient Safety (PIPS) committee is a requirement of the CSV program.

   i. This committee must meet at least quarterly but may meet as often as a center determines it is necessary. Our PIPS committee meets monthly for 1 h.
   ii. This committee should be comprised of a representative from each children's surgical subspecialty, and these members are required to uphold a 50% attendance rate. This also includes procedural specialists from Gastroenterology and Pulmonology.
   iii. The Medical Director of the Children's Surgery program, the Program Manager, and the Medical Director of Anesthesia are required to uphold a 75% attendance rate.
   iv. Developing a PIPS committee requires buy-in from all specialties and participation by all involved to create a robust group and discussion.
   v. Utilization of presentation slides of meeting items is helpful to keep meeting on track and consistent.
   vi. PIPS meetings can occur using socially distant methods via web conference if needed.
   vii. Sample PIPS committee agenda consists of:

      a. Call to order/attendance
      b. Children's OR committee meeting
      c. New business
      d. CSV updates
      e. Emergent/urgent case review
      f. Transfer, over/under triage review
      g. Performance improvement projects
      h. Performance improvement and patient safety review

         a. Appendix 2 data review

            i. Surgical case volume
            ii. Appendix 2 events

         b. NSQIP occurrence review

      i. Peer review/multidisciplinary discussion

   viii. Peer review cases should be assessed for:

      a. Progression of disease, known complication, deviation from accepted standard of care
      b. Contributing factors—patient related, provider, related, system related, or none

   ix. Mortality reviews should be classified as:

      a. Anticipated with improvement opportunity
      b. Anticipated without improvement opportunity

        c. Unanticipated with improvement opportunity
        d. Unanticipated without improvement opportunity

    x. Actions from multidisciplinary peer review cases could be:

        a. Discussion at Morbidity and Mortality Conference (M&M) (date/time)
        b. No action required
        c. Policy revision/adoption recommended
        d. Referral to peer review
        e. Referral for further review to another department/group
        f. Education of service/individual/institute
        g. Multidisciplinary review
        h. Others

3. Utilization of locums tenens surgeons in pediatric general surgery—locum tenens comes from Latin “to hold the place, to substitute for.”

    a. The demand for pediatric surgeon outpaces the supply in many communities in the United States. This leads to a shortage of available surgeons. Often this shortfall is worst in rural areas.
    b. Many treatment facilities have relied on the employment of locum tenens pediatric surgeons to maintain uninterrupted surgical care for children.
    c. Guidelines must be followed to ensure that the use of visiting surgeons does not affect the quality and continuity of care provided.
    d. Proposed standards for the use of locum tenens pediatric surgeons were published in the *Journal of Pediatric Surgery* in 2013 [4].

        i. High standards of patient care must be maintained.

            1. Locum tenens pediatric surgeons should be board certified or eligible in pediatric surgery.
            2. They should maintain professional membership.
            3. All local credentialing requirements must be met.
            4. A local physician extender should assist the visiting surgeon in coordinating care and reaching multidisciplinary services.
            5. Uninterrupted service should be ensured with an established pediatric surgical practice in place in the institution which can provide postoperative recovery for children as well as long-term care.
            6. Pediatric surgery should not be provided at an institution solely by rotating locum surgeons. There must be a full-time staff member who provides the continuity of care necessary for safety.

        ii. The visiting surgeon should continue educational activities for residents and students.
        iii. Arrangements must be made for clear communication to and from the visiting surgeon.

            1. A hospital representative must provide a detailed orientation to a new surgeon to ensure that they can find the emergency department,

wards, operating room, and clinic. The surgeon should be given contact information for all resources they may need to provide surgical care to children.

2. There needs to be a detailed transfer of care directly between surgeons when the locum tenens pediatric surgeon arrives and departs to be sure that continuity of care is maintained for children on the pediatric surgical service.

iv. Although it is always a risk of discontinuity to utilize visiting surgeons, precautions can be taken to minimize the effect on children. Every institution providing pediatric surgical care should have at least one full-time pediatric surgeon to maintain practices and standards. When possible, the same locum tenens pediatric surgeons should be employed to reduce the total variability in care.

v. All institutions providing pediatric surgical care should have pre-established protocols of transfer of children to elevated level of care when needed. The need to transfer may vary depending on not only local resources available but also the judgment and decision of the visiting locum tenens pediatric surgeon (Tables 1 and 2).

**Table 1** Requirements for Level 1 and Level 2 children's surgical centers

| Requirement | Children's Level I | Children's Level II | Children's Level II (neonatal scope of service only) |
|---|---|---|---|
| Freestanding children's hospital or comprehensive pediatric care unit within general hospital organization | E | E | – |
| Children's surgical service | E | E | E |
| Pediatric surgeons | E | E | E |
| Pediatric anesthesiologists | E | E | E |
| Pediatric emergency medicine physicians | E | D | – |
| Pediatric radiologists | E | E | E |
| Other surgical specialists | E | E | E (pediatric ophthalmology) |
| Pediatric rapid response team | E | E | E |
| Surgery-specific children's continuing medical education for children's surgery medical director and liaisons | E | E | E |
| Neonatal intensive care unit | Level IV | Level III or greater | Level III or greater |
| Neonatologists | E | E | E |
| Pediatric emergency department | E | E | – |
| Pediatric intensive care unit | E | D | – |

**Table 1** (continued)

| Requirement | Children's Level I | Children's Level II | Children's Level II (neonatal scope of service only) |
|---|---|---|---|
| Pediatric critical care medicine physicians | E | E | – |
| Pediatric acute care unit | E | E | – |
| Pediatric resuscitation equipment in all appropriate patient-care areas | E | E | E |
| Children's surgical program manager or coordinator | E | E | E |
| Surgical data collection | E | E | E |
| Child-life and family-support programs | E | E | E |
| Pediatric social work/child protective services | E | E | E/D |
| Community outreach programs | E | E | – |
| Children's education programs | E | E | – |
| Surgical research | E | D | D |
| Minimum number of annual surgical procedures for children younger than 18 years | 1000 | – | – |
| Children's surgical performance improvement and patient safety program | E | E | E |

Source: Optimal Resources for Children's Surgical Care v.1 [5]. (Note: Case number requirements changed with 2021 update)
Abbreviations: D, desired; E, essential

**Table 2** Summary of children's surgical center standards with expected scope of practice

| | Level 1 | Level II | Level III | Specialty musculoskeletal | Specialty oncology |
|---|---|---|---|---|---|
| Age | Any | Any | >6 months | Any | Any |
| ASA | 1–5 | 1–3[a] | 1–2 | 1–5 | 1–5 |
| Co-Morbidities | All—complex | Typically, single or multiple specialty management | None—healthy children | All—complex | All—complex |
| Operations[b] | All—complex diseases, multispecialty care | Common anomalies, single specialty or multiple specialty care | Common "low-risk" procedures by single specialty | All—complex musculoskeletal diseases, multispecialty care | All—complex oncology-related diseases, multispecialty care |

(continued)

**Table 2** (continued)

| | Level 1 | Level II | Level III | Specialty musculoskeletal | Specialty oncology |
|---|---|---|---|---|---|
| Ambulatory[c] | ASA 1–3, guidelines for post-anesthesia monitoring | ASA 1–3, guidelines for post anesthesia monitoring | Age >6 months, Healthy children | ASA 1–3, guidelines for post-anesthesia monitoring | ASA 1–3, guidelines for post-anesthesia monitoring |

Source: Optimal resources for Children's Surgical care: 2021 Standards [3]

[a] Emergent procedures in some patients of ASA >3 may be appropriate in neonatal patients, such as those with necrotizing enterocolitis

Infants and children who have emergent or life-threatening surgical needs and cannot be reasonably delayed for transport should receive initial stabilization and necessary care at the site of presentation

[b] For neonates, types of anomalies and diseases that should have pediatric subspecialty care are further delineated in American Academy of Pediatrics. Levels of neonatal care: Committee on Fetus and Newborn. Pediatrics. 2012;130(3):587–597. Depending on patient age, comorbidities, and need for a multidisciplinary surgical approach, these operations may be appropriate for either Level I or Level II centers

[c] Ambulatory sites of care must be included when the ambulatory site is an integrated site of the parent institution and is served by the same medical staff in these recommended levels of institutional designation. The site of care may be physically attached to, or integrated into, the hospital and must be a component of a demonstrably integrated children's health care delivery system that provides these defined resources

Abbreviations: ASA, American Society of Anesthesiologists perioperative risk score

## References

1. American College of Surgeons National Surgical Quality Improvement Program Pediatric. Chicago, IL: American College of Surgeons; 2021. https://www.facs.org/quality-programs/childrens-surgery/pediatric. Accessed 12 Aug 2021.
2. Task Force for Children's Surgical Care. Optimal resources for children's surgical care in the United States. J Am Coll Surg. 2014;218(3):479–87.
3. Optimal Resources for Children's Surgical Care: 2021 Standards. Chicago, IL: American College of Surgeons; 2021. https://www.facs.org/-/media/files/quality-programs/csv/2021_optimal_resources_for_childrens_surgical_care.ashx. Accessed 11 Aug 2021.
4. Loe WA, Guzzetta PC, Lessin MS, Nakayama DK. Proposed standards for use of locum tenens coverage in pediatric surgery practices. Corres J Ped Surg. 2013;48:700–3.
5. Optimal Resources for Children's Surgical Care v.1. Chicago, IL: American College of Surgeons; 2015. https://www.facs.org/-/media/files/quality-programs/csv/acs-csv_standards-manual.ashx. Accessed 12 Aug 2021.

## *Further Reading*

Approved by Board of Regents of the American College of Surgeons. Statement on maintaining surgical access with a locum tenens surgeon. Bull Am Coll Surg. 2018;103(1):44–6.

Children's Surgery Verification Quality Improvement Program. Chicago, IL: American College of Surgeons; 2021. https://www.facs.org/childrensverification. Accessed 11 Aug 2021.

# COVID-19 Pandemic and Contamination Response

**Max D. Hazeltine and Michael P. Hirsh**

## Origins

Scientists agree that a novel coronavirus, severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), unlike any seen in humans previously, made its way into the general population of Wuhan, China, sometime in late 2019. This type of virus has been found in bats and pangolins, and there is a popular wet market in Wuhan where people can buy these animals for home preparation as foodstuff. Ingestion of contaminated food or proximity to these animals could have been the origin of patient zero. Wuhan is also the location of a virology institute that, among other projects, had been studying coronavirus in these two animal species. Another theory regarding the origin of COVID-19 was that a lab accident or the infection of a lab worker was the inciting event; however, studies have deemed this unlikely.

The initial epidemic spread rapidly before the Chinese government notified international medical agencies. Those who attempted to alert the international community on social media were suppressed. The lack of transparency caused a devastating loss of preparation time. The Chinese government did not formally acknowledge the existence of the epidemic until early January 2020.

By January 13, 2020, there was the first confirmed case of the new virus outside of China, in a 61-year-old woman in Thailand. The Chinese government did not acknowledge that the novel coronavirus was spreading by human contact until January 20, 2020. The following day, the United States (US) reported its first

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confirmed case in Washington state. By January 31 the Trump administration restricted travel from China. Within the next few weeks, cases would be reported in six states within the United States, but also internationally in France, Italy, Iran, and Brazil. Case numbers continued to mount worldwide until finally the World Health Organization declared the disease a pandemic on March 11, 2020.

The US Health and Human Services Secretary declared a national state of emergency on January 31, 2020. In 3 months between the initial recognition of the epidemic in China and the Center for Disease Control and Prevention (CDC) declaration, preparations locally in the United States and in countries around the world were delayed and varied in terms of levels of severity.

The US Public Health Service, which had long been under-resourced and understaffed, now faced the daunting task of trying to mitigate the spread and educate the populace about a virus with little information available about the nature of the virus itself, nor the pathophysiology of any resultant disease inflicted. Further complicating these efforts was significant politicization of the response to the disease from the highest levels of US government.

## Pathophysiology and Clinical Presentation

SARS-CoV-2, the virus that causes coronavirus disease 2019 (COVID-19), spreads by respiratory droplets and binds to the angiotensin-converting enzyme-2 (ACE2) receptor and most often affects the airway. Once there, it causes a variable immune response that seems to be age-related, as the expression of the ACE2 increases with age. It most impacted those who were unable to distance from family, workers, and schoolmates. There was great concern in the early days of the pandemic as to how much spread was done by contact on surfaces. However, frequent handwashing or hand sanitizer use seemed to limit this source of spread. Another initial challenge was determining whether the virus spread solely by droplets or via aerosolized particles, and it remains unclear how significantly aerosol spread contributes to transmission. The challenge for healthcare workers from first responders to intensive care unit physicians was the requirement to don personal protective equipment to protect themselves from being infected with the virus themselves. Powered air-purifying respiratory (PAPR) devices or N95 masks as well as eye protection, gloves, and gowns were essential for healthcare workers but in the first surge were in short supply leading to a great deal of viral spread among medical staff.

The clinical presentation of COVID-19 is classically a dry, nonproductive cough, fever, headache, body aches, and very importantly loss of the sense of smell and taste. Furthermore, the virus has prothrombotic properties that can lead to thromboembolic events such as deep venous thrombosis and pulmonary emboli. Gastrointestinal complaints such as nausea, diarrhea, and abdominal pain are also common. Unique entities like COVID toes that seem to stem from microvasculature thrombosis causing pain and discoloration have also been described.

Several risk factors have been identified, such as age over 64 years, hypertension, diabetes, chronic obstructive pulmonary disease, cardiovascular disease, and cerebrovascular disease. Younger patients frequently have minimal or no symptoms. Even though younger patients tend to not exhibit as severe a clinical presentation as older patients, younger patients have developed significant sequelae such as myocarditis, acute kidney injury, or hepatic dysfunction. Additionally, there is a phenomenon now being observed 1 year into the pandemic of patients who have coalesced from their initial COVID-19 infection returning with what is called long-COVID or COVID long-hauler disease: persistent and debilitating symptoms that are heterogenous in nature and etiology. It is unclear what are best treatments and how long these manifestations will last.

In children an illness called multisystem inflammatory syndrome in children (MIS-C) has emerged. It is rare (<1% of children with confirmed COVID-19) and believed to be due to immune dysregulation after the acute infection has resolved. Presenting symptoms are typically fever, abdominal pain, vomiting, diarrhea, rash, and conjunctivitis, but can progress to multiorgan system failure, including shock, myocardial dysfunction, arrhythmia, acute kidney injury, hepatitis, as well as acute respiratory failure. The overall mortality is approximately 1–2%; however, data are limited.

## Management

Initial approaches to COVID-19 management borrowed existing knowledge from acute respiratory distress syndrome given the novelty of COVID-19. These included supportive respiratory care with oxygen, judicious fluid management, and early intubation and mechanical respiratory support. For critically ill patients who were already on mechanical ventilation, prone positioning and paralysis were utilized to improve oxygenation. However, it gradually became apparent that once intubated many patients plateaued and had difficulty weaning from mechanical ventilation. Therefore, prone positioning prior to intubation, high flow nasal cannula, and noninvasive positive pressure ventilation have been utilized to prevent disease progression, although high flow nasal cannula and noninvasive positive pressure ventilation are controversial given the concern for further aerosolization of viral particles.

As the pandemic progressed, a number of therapies have been tested and proven to be effective in COVID-19 management. Systemic glucocorticoids have shown a mortality benefit in critically ill patients receiving mechanical ventilation or oxygen therapy. Additionally, remdesivir, an RNA polymerase inhibitor that inhibits SARS-CoV-2 replication, may reduce time to recovery. Finally, ongoing trials are exploring the role of convalescent plasma and monoclonal antibody therapy.

From a surgical perspective, there were a number of issues with the perioperative management of patients with COVID. A major concern was how we could operate safely and prevent infection of healthcare workers. Some institutions required providers wear a PAPR or an N95 respirator, or sometimes both. Intubation was often done with both a PAPR and N95, only by the most experienced anesthesiologist available, and while all other providers were outside the operating room. Once testing was readily available, institutional guidelines varied significantly in terms of the timing of a COVID test preoperatively. Cases that could be delayed often were, and in some instances, such as acute appendicitis, managed non-operatively to avoid the risk of aerosolization of virus. Institutional responses were highly variable, and providing proper care safely was a challenge. Fortunately, a number of studies in adults that examined risk of COVID-19 and healthcare workers found minimal risk of contracting COVID-19 when using appropriate PPE (masks and gloves) as well as hand hygiene. Additionally, perioperative risk of COVID-19 appears to be quite low even with high community rates.

## Mitigation

Mitigation of viral spread, as most are likely aware, involves the three W's: (1) Wear a mask, (2) Wary of distance (i.e., social distance), and (3) Wash your hands. Further public health measures of locking down communities, closing on-site retail and dining opportunities, as well as modifying school and college to remote or hybrid models were other measures that have been implemented to prevent the spread of COVID-19.

COVID-19 vaccines, which were mRNA vaccines encoding the prefusion spike glycoprotein of SARS-CoV-2, rather than an attenuated live virus, were introduced in clinical trials by August 2020 and in emergency use by January 2021. mRNA is the intermediary step from DNA, the genetic code for proteins, to the protein creation itself. The mRNA encodes for a particular protein, and the use of mRNA vaccines is advantageous for several reasons:

1. Safety: mRNA is non-infectious, does not integrate into the host DNA, and is degraded by normal cellular processes.
2. Efficacy: modifications can make mRNA more stable and translatable, and mRNA can be delivered easily without significant host degradation.
3. Production: mRNA vaccines can be rapidly and inexpensively produced.

The development of mRNA therapies actually began in 1990 and had numerous clinical trials under investigation years before the COVID-19 pandemic. In fact, ModernaTX, Inc. had funded trials of an influenza mRNA vaccine starting in 2015. These prior successes were essential to having the proper infrastructure and knowledge in order to rapidly develop a COVID-19 vaccine. Additionally, significant public-private partnerships and investments were made to facilitate candidate vaccine production, leading to faster vaccine production than previously experienced. There is a good probability that by year-end 2021 we will have achieved herd immunity in the United States, as over 330 million vaccine doses have been administered as of July 6, 2021 (approximately 67% of the adult population).

COVID-19 in the meantime did not fail to follow in the footsteps of epidemic precursors such as SARS and MERS in developing mutations that made them more transmissible. As of July 2021, four variants of concern have been identified by the CDC in the United States. Studies suggest that these variants are recognized by antibodies generated from vaccination, although these variants seem to be more transmissible which may lead to further spread of COVID-19. These variants continue to present themselves and herald the possibility that we will need to develop boosters to continue our pursuit of herd immunity.

## Impact of the COVID-19 Pandemic on Pediatric Surgery

COVID-19 took a tremendous toll on the United States and the world. Since the onset of the pandemic, over 600,000 Americans have died. Since all elective surgery was shut down for the greater part of 2020, many parents have put off "elective" procedures like hernia, hydrocele, and pectus excavatum repairs. The long-term outcomes from these delays are not entirely known, but evidence is emerging that delays have led to higher rates of acute appendicitis and severe diabetic ketoacidosis and likely other morbidities that have yet to be reported. Another major concern that has been raised is the impact on surgical trainees. One study found significant impacts of the pandemic on learner experience, with reductions in clinical experiences as well as didactic education. More concerning were findings regarding worse well-being of learners.

The four most salient pediatric surgery issues that the COVID-19 pandemic has raised are:

1. There will be a 2-year period where surgical trainees have not been properly exposed to the number of pediatric surgery cases to either qualify themselves to be pediatric surgeons or give them enough exposure to decide to pursue the specialty.
2. The psychological effects of the "virtual" school years and lack of social bonding will undoubtedly cause some (hopefully) reparable harm to the development of this "COVID generation."
3. The pandemic exposed issues with health disparities that all parts of the medical machinery were already realizing needed addressing before the pandemic. Minority and socioeconomic communities clearly fared worse than White or more economically advantaged communities. These systemic issues will have to be aggressively addressed moving forward.
4. The US public health system needs to be underwritten in a more robust way as this type of outbreak may become more frequent given the issues of climate change, as well as ongoing widening of economic inequality and racial disparities.

## Conclusion

The COVID-19 pandemic took a devastating toll on the worldwide community. The pediatric population, although not directly impacted by the virus as greatly as adult or elderly communities, also was impacted by severe disease, as evidenced by MIS-C. However, more insidious or long-term sequelae from the pandemic have yet to be fully uncovered. The harm to children from delays in care is likely substantial, and the impact on children's mental health and development may not be fully known for decades. Finally, the pandemic may have detrimental effects on surgical trainees as many missed opportunities to perform pediatric surgery procedures or have exposure to the field.

## Study Questions

1. A 10-year-old male is presenting to the emergency room with 3 days of fevers, right lower quadrant abdominal pain, nausea, and diarrhea. The patient's older brother recovered from COVID-19 6 weeks prior. His vitals are notable for a temperature of 38.4 °C and heart rate of 115. On exam his abdomen is mildly tender in the lower abdomen, but without rebound or guarding. He was found to be mildly anemic, with lymphocytopenia. An ultrasound of the abdomen did not demonstrate any intra-abdominal pathology but diffuse reactive lymphadenopathy. What is the most likely diagnosis in this patient?

    a. COVID-19
    b. Multisystem inflammatory syndrome in children (MIS-C)
    c. Kawasaki disease
    d. Gastroenteritis

    Answer: (b) Multisystem inflammatory syndrome in children (MIS-C). This patient is presenting with fevers and abdominal pain in the context of a recent exposure to COVID-19. He likely had an asymptomatic COVID-19 infection and coalesced but now is presenting with signs of systemic inflammation and a normal ultrasound. This constellation of findings is most consistent with MIS-C, which is thought to be due to immune dysregulation after COVID-19 infection. Management is primarily supportive, with data continuing to emerge regarding best management practices.

## Further Reading

Association AH. U.S. declares coronavirus a public health emergency, CDC updates guidance. 2020 January 31, 2020.

Bethell GS, Rees CM, Sutcliffe JR, Hall NJ. Management and early outcomes of children with appendicitis in the UK and Ireland during the COVID-19 pandemic: a survey of surgeons and observational study. BMJ Paediatr Open. 2020;4(1):e000831.

Boghani P. A timeline of China's response in the first days of COVID-19. Frontline PBS. 2021 February 2, 2021.

Chen P, Nirula A, Heller B, Gottlieb RL, Boscia J, Morris J, et al. SARS-CoV-2 neutralizing antibody LY-CoV555 in outpatients with Covid-19. N Engl J Med. 2021;384(3):229–37.

Chu DK, Akl EA, Duda S, Solo K, Yaacoub S, Schünemann HJ. Physical distancing, face masks, and eye protection to prevent person-to-person transmission of SARS-CoV-2 and COVID-19: a systematic review and meta-analysis. Lancet. 2020;395(10242):1973–87.

Dickenson EJ, Jordan RW, Poole C, Shyamalan G, Arbuthnot J, Makrides P, et al. Is it safe to perform elective orthopaedic surgery with high community rates of COVID-19? An observational cohort study. Ann R Coll Surg Engl. 2021 Jun;103(6):415–9.

Dufort EM, Koumans EH, Chow EJ, Rosenthal EM, Muse A, Rowlands J, et al. Multisystem inflammatory syndrome in children in New York State. N Engl J Med. 2020;383(4):347–58.

Ellison EC, Spanknebel K, Stain SC, Shabahang MM, Matthews JB, Debas HT, et al. Impact of the COVID-19 pandemic on surgical training and learner well-being: report of a survey of general surgery and other surgical specialty educators. J Am Coll Surg. 2020;231(6):613–26.

Feldman RA, Fuhr R, Smolenov I, Mick Ribeiro A, Panther L, Watson M, et al. mRNA vaccines against H10N8 and H7N9 influenza viruses of pandemic potential are immunogenic and well tolerated in healthy adults in phase 1 randomized clinical trials. Vaccine. 2019;37(25):3326–34.

Freeman EE, McMahon DE, Lipoff JB, Rosenbach M, Kovarik C, Desai SR, et al. The spectrum of COVID-19-associated dermatologic manifestations: an international registry of 716 patients from 31 countries. J Am Acad Dermatol. 2020;83(4):1118–29.

Gautret P, Million M, Jarrot PA, Camoin-Jau L, Colson P, Fenollar F, et al. Natural history of COVID-19 and therapeutic options. Expert Rev Clin Immunol. 2020;16(12):1159–84.

Han HJ, Nwagwu C, Anyim O, Ekweremadu C, Kim S. COVID-19 and cancer: from basic mechanisms to vaccine development using nanotechnology. Int Immunopharmacol. 2021;90:107247.

Health NIo. Coronavirus disease 2019 (COVID-19) treatment guidelines. National Institutes of Health; 2020 April 21, 2021.

Ho RJY. Warp-speed Covid-19 vaccine development: beneficiaries of maturation in biopharmaceutical technologies and public-private partnerships. J Pharm Sci. 2021;110(2):615–8.

Horby P, Lim WS, Emberson JR, Mafham M, Bell JL, Linsell L, et al. Dexamethasone in hospitalized patients with Covid-19. N Engl J Med. 2021;384(8):693–704.

J. P. Scientists are working on booster shots in case COVID-19 vaccines lose their effect. National Public Radio. 2021. April 22, 2021.

Li X, Giorgi EE, Marichannegowda MH, Foley B, Xiao C, Kong XP, et al. Emergence of SARS-CoV-2 through recombination and strong purifying selection. Sci Adv. 2020;6(27) https://doi.org/10.1126/sciadv.abb9153.

Liu YC, Kuo RL, Shih SR. COVID-19: the first documented coronavirus pandemic in history. Biom J. 2020;43(4):328–33.

McCreary EK, Angus DC. Efficacy of remdesivir in COVID-19. JAMA. 2020;324(11):1041–2.

Medicine Johns Hopkins University. Coronavirus resource center. Johns Hopkins University & Medicine; 2021.

Morens DM, Breman JG, Calisher CH, Doherty PC, Hahn BH, Keusch GT, et al. The origin of COVID-19 and why it matters. Am J Trop Med Hyg. 2020;103(3):955–9.

Nakra NA, Blumberg DA, Herrera-Guerra A, Lakshminrusimha S. Multi-system inflammatory syndrome in children (MIS-C) following SARS-CoV-2 infection: review of clinical presentation: Hypothetical Pathogenesis, and Proposed Management. Children (Basel). 2020;7(7):69.

Nishimura T, Uwamino Y, Uno S, Kashimura S, Shiraki T, Kurafuji T, et al. SARS-CoV-2 infection among medical institution faculty and healthcare workers in Tokyo, Japan. Intern Med. Jun 19; 2021.

Oliver SE, Gargano JW, Marin M, Wallace M, Curran KG, Chamberland M, et al. The advisory committee on immunization practices' interim recommendation for use of Pfizer-BioNTech COVID-19 Vaccine—United States, December 2020. MMWR Morb Mortal Wkly Rep. 2020;69(50):1922–4.

Organization WH. Archived: WHO timeline—COVID-19. 2020 April 27, 2020.

Oualha M, Bendavid M, Berteloot L, Corsia A, Lesage F, Vedrenne M, et al. Severe and fatal forms of COVID-19 in children. Arch Pediatr. 2020;27(5):235–8.

Pan Z, Zhang H, Yang J, Tang S, Cheng Z, Wu K, et al. Surgical masks for protection of health care personnel against Covid-19: results from an observational study. Clin Invest Med. 2021;44(2):E48–54.

Pardi N, Hogan MJ, Porter FW, Weissman D. mRNA vaccines—a new era in vaccinology. Nat Rev Drug Discov. 2018;17(4):261–79.

Perego E, Callard F, Stras L, Melville-Jóhannesson B, Pope R, Alwan NA. Why the patient-made term 'long covid' is needed. Wellcome Open Res. 2020;5:224.

Piechotta V, Chai KL, Valk SJ, Doree C, Monsef I, Wood EM, et al. Convalescent plasma or hyperimmune immunoglobulin for people with COVID-19: a living systematic review. Cochrane Database Syst Rev. 2020;7(7):Cd013600.

Polack FP, Thomas SJ, Kitchin N, Absalon J, Gurtman A, Lockhart S, et al. Safety and efficacy of the BNT162b2 mRNA Covid-19 vaccine. N Engl J Med. 2020;383(27):2603–15.

Prevention CfDCa. About variants of the virus that causes COVID-19. Centers for Disease Control and Prevention; 2021. April 2, 2021a.

Prevention CfDCa. COVID-19 vaccinations in the United States. Centers for Disease Control and Prevention; 2021. April 27, 2021b.

Rabbone I, Schiaffini R, Cherubini V, Maffeis C, Scaramuzza A. Has COVID-19 delayed the diagnosis and worsened the presentation of Type 1 diabetes in children? Diabetes Care. 2020;43(11):2870–2.

Ranney ML, Griffeth V, Jha AK. Critical supply shortages: the need for ventilators and personal protective equipment during the COVID-19 pandemic. N Engl J Med. 2020;382(18):e41.

Romero-Velez G, Rodriguez Quintero JH, Pereira X, Nussbaum JE, McAuliffe JC. SARS-CoV-2 during abdominal operations: are surgeons at risk? Surg Laparosc Endosc Percutan Tech. 2021;31:674–8.

Singh AK, Singh A, Singh R, Misra A. Remdesivir in COVID-19: a critical review of pharmacology, pre-clinical and clinical studies. Diabetes Metab Syndr. 2020;14(4):641–8.

Snapiri O, Rosenberg Danziger C, Krause I, Kravarusic D, Yulevich A, Balla U, et al. Delayed diagnosis of paediatric appendicitis during the COVID-19 pandemic. Acta Paediatr. 2020;109(8):1672–6.

Stasi C, Fallani S, Voller F, Silvestri C. Treatment for COVID-19: an overview. Eur J Pharmacol. 2020;889:173644.

Taylor DB. A timeline of the coronavirus pandemic. New York Times. 2021 March 17, 2021.

Webb Hooper M, Nápoles AM, Pérez-Stable EJ. COVID-19 and racial/ethnic disparities. JAMA. 2020;323(24):2466–7.

Wiersinga WJ, Rhodes A, Cheng AC, Peacock SJ, Prescott HC. Pathophysiology, transmission, diagnosis, and treatment of coronavirus disease 2019 (COVID-19): a review. JAMA. 2020;324(8):782–93.

Yee S, Tan CS, Khan A, Lee KS, Goh BH, Ming LC. SARS-COV-2 as an artificial creation: scientific arguments and counterarguments. J Med Life. 2021;14(1):118–20.

# Part II
# Perioperative Issues

# Fluids and Electrolytes

**Ronald J. Scorpio and Alfred P. Kennedy Jr**

**Key Points**

1) Children are not small adults. Babies are not small children. This is particularly true in the management of fluids in electrolytes of the pediatric surgical patient.
2) As children grow, the relative size of body fluid compartments changes with a decreasing size of the extracellular fluid compartment and an increase in the non-fluid compartment.
3) Urine output must be followed in critically ill children to track their fluid status.
4) To properly treat electrolyte abnormalities, the underlying cause must be identified and addressed.

1. Newborns
    A. Term newborns have decreased glomerular filtration rate (GFR). Term newborns are capable of concentrating urine to an osmolarity of 800 mOsm/kg, whereas preterm infants are capable of only 600 mOsm/kg (contrast this to an adult nephron which is capable of 1200 mOsm/kg). Initially after birth, infants exhibit an obligatory diuretic phase during this transitional period.

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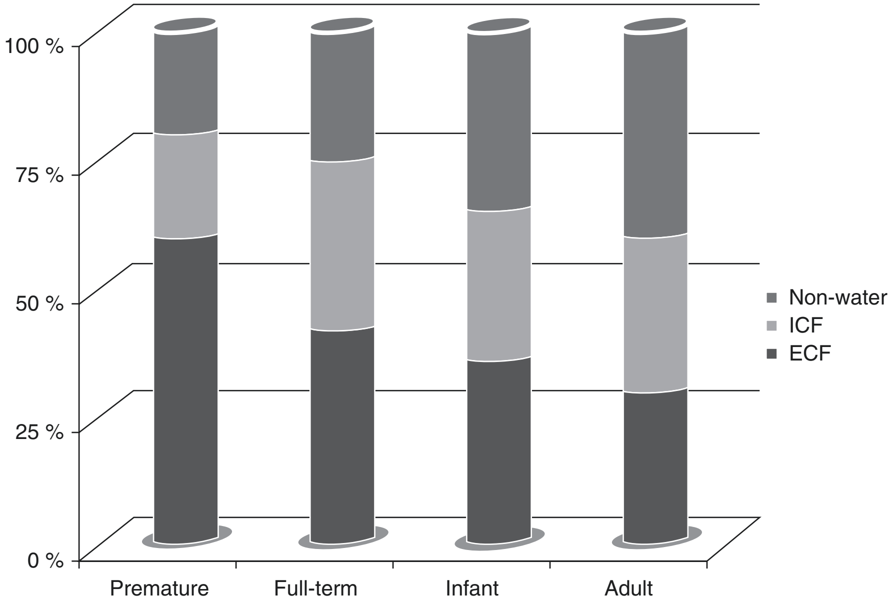

**Fig. 1** Relative body fluid compartments by age. As children grow, the relative size of body fluid compartments changes (Source: Christopher Coppola). *ECF* extracellular fluid, *ICF* intracellular fluid

B. The distal convoluting tubule in infants is relatively unresponsive to antidiuretic hormone (ADH) secretion.
C. Newborns are unable to excrete excess sodium (Na), and premature infants are considered salt wasters due to the immaturity of the renal tubule. However, both populations can excrete relatively large quantities of dilute urine. Large solute loads are unnecessary and may be deleterious.
D. Consequently, there are major shifts in the water content of the intracellular and extracellular compartments and total body water over the first year of life. Total blood volume is relatively higher in an infant (90 mL/kg) compared to the adult (70 mL/kg) (Fig. 1).

2. Urine output:

   A. Measurement of hourly urine output is essential in critically ill children.
   B. Catheterization of small infants, especially the male, is fraught with complication and should only be undertaken by experienced personnel.
   C. In order to excrete adequate solute load, the following are approximations of adequate hourly urine output:

      i. Infant ~ 2 mL/kg
      ii. Child ~ 1 mL/kg
      iii. Adolescent ~ 0.5 mL/kg

D. These are only approximate volumes and are certainly an underestimate in the critically ill child. Reassessment of the clinical situation and the patient's hydration status is vital to success of resuscitation (Tables 1 and 2).
E. Other signs of adequate perfusion include capillary refill, skin temperature, heart rate, level of consciousness, and fontanel examination in the infant.

3. Fluid requirements: "4–2–1 rule."

A. 4 mL/kg/h for the first 10 kg of dry bodyweight.
B. 2 mL/kg/h for the next 10 kg of dry bodyweight.
C. 1 mL/kg/h for each additional kg of dry bodyweight.
D. Example: A 20 kg child will need approximately 60 mL/h of maintenance fluid.
E. This is only an estimate and may require frequent adjustment depending on urine output, heart rate, and other signs of perfusion.

4. Replacement therapy:
Three basic aspects need to be considered:

A. Maintenance fluid requirements
B. Existing deficit

1. Calculated from bodyweight based upon degree of dehydration:

**Table 1** Compositions of common intravenous fluids

| Fluid | Na (mEq/L) | Cl (mEq/L) | K (mEq/L) | Lactate (mEq/L) | Glucose (g/L) | Osmolarity |
|---|---|---|---|---|---|---|
| Ringer's lactate | 130 | 109 | 4 | 28 | 0 | 272 |
| Normal saline | 154 | 154 | 0 | 0 | 0 | 308 |
| ½ normal saline | 77 | 77 | 0 | 0 | 0 | 154 |
| D5W | 0 | 0 | 0 | 0 | 50 | 252 |
| D10W | 0 | 0 | 0 | 0 | 100 | 505 |
| 3% saline | 513 | 513 | 0 | 0 | 0 | 1026 |

**Table 2** Osmolar load and ideal renal water

| Diagnosis | Osmolar load (mOsm/kg/24 h) | Ideal renal water (mL/kg/h) |
|---|---|---|
| Bowel atresia | 14.7 ± 6.7 | 2.2 ± 0.95 |
| Esophageal atresia | 11.6 ± 3.0 | 1.7 ± 0.4 |
| Gastroschisis | 11.7 ± 8.6 | 1.7 ± 0.7 |
| Necrotizing enterocolitis | 17.75 ± 8.6 | 2.6 ± 1.25 |

Adapted from O'Neill JA, Fonkalsrud EW, Coran AG, Calmadone AA. Principles of Pediatric Surgery 2nd Ed Mosby 1995

Decreased peripheral perfusion (capillary refill)
Acidotic (deep) breathing
Decrease skin turgor
Increase thirst

2. 0–4% loss of bodyweight, no clinical signs

   4–10% loss of bodyweight, clinical signs are present.
   >10% loss bodyweight, peripheral circulatory failure.

C. Continuing losses during therapy

   Important to have an accurate recording of fluid balance, ins and outs, and the type of lost: gastric, diarrhea, fistula output, or other losses. This will determine the type of replacement fluid. During a laparotomy, for instance, up to 10 mL/kg/h may be lost while the abdomen is open.

5. Insensible water loss:
   A. Respiratory: May be mitigated by humidifying inspired gases in those children who require mechanical ventilation.
   B. Skin.
      i. Especially significant in premature infants.
      ii. Mitigated by wrapping extremities, humidified incubators.

Common electrolyte anomalies:

6. Sodium (Na): Newborn requires approximately 2 mEq/kg/day.
   A. Hyponatremia
      i. Most common electrolyte abnormality in surgical patients.
      ii. Usually indicates hypo-osmolality and excess extracellular water.
      iii. Symptoms are CNS related and include headache, nausea, lethargy, hallucinations, and coma.
      iv. Common causes in children:
         a. Excess ADH secretion in postoperative period
         b. Bowel obstruction
         c. Peritonitis
         d. Renal impairment
         e. Hypotonic intravenous fluid administration
         f. Diuretics
         g. Mannitol infusion
      v. Correction requires treatment of underlying cause and slow adjustment of serum sodium to prevent central pontine myelinolysis.
      vi. Measurement of serum osmolality can assist the clinician to codify three clinically different forms of hyponatremia: hyper-, iso-, and hypo-osmolar hyponatremia.

vii. Furthermore, hypo-osmolar hyponatremia may be further classified by clinical hydration status.
viii. Hypertonic hyponatremia is primarily a manifestation of hyperglycemia or infusion of hypertonic intravenous solutions such as mannitol.
ix. Isotonic hyponatremia is seen in the state of hyperlipidemia or hyperproteinemia.
x. Isovolumic, hypotonic hyponatremia is exclusively seen in the setting of the syndrome of inappropriate antidiuretic hormone secretion (SIADH). Oftentimes, this is a byproduct of a severe closed head injury with increased intracranial pressure.
xi. Hypovolemic, hypotonic hyponatremia is usually seen with excessive gastrointestinal or renal losses or "third-spacing" after trauma or surgery. Adrenal insufficiency may be a culprit as well.
xii. Hypervolemic, hypotonic hyponatremia is seen in the face of congestive heart failure, cirrhosis, or nephrotic syndrome.

B. Hypernatremia

   i. Hypernatremia may also be classified as to the clinical volume status of the patient.
   ii. Causes of hypervolemic hypernatremia are usually iatrogenic crystalloid infusion. Excess mineralocorticoids may also produce this effect.
   iii. Diabetes insipidus produces a relative isovolemic hypernatremia.
   iv. Hypovolemic hypernatremia is a manifestation of excessive insensible losses, diuretic therapy, renal insufficiency, glycosuria, or adrenal insufficiency.

7. Potassium (K):

   A. As most potassium is intracellular, serum potassium correlates poorly with the amount of total body potassium.
   B. Daily requirement is 1–2 mEq/kg.
   C. Potassium infusion should be undertaken only in the face of normal urine output.
   D. Hypokalemia:

      i. Symptoms include muscle weakness, cramps, myalgia, and paralysis. Common causes in children include:

         a. Diarrhea
         b. Diuretics (except potassium sparing, e.g., spironolactone)
         c. Excessive vomiting (e.g., pyloric stenosis)
         d. Metabolic alkalosis
         e. Prolonged nasogastric tube aspiration
         f. Inadequate K administration (TPN)

      ii. Treatment requires recognition of underlying cause and most commonly K supplementation either orally or intravenously. When the patient is symptomatic (e.g., arrhythmia), rapid repletion requires placement of a central line. No more than 20 mEq/L should be given via a peripheral IV.

E. Hyperkalemia:

   i. Symptoms depend on serum levels of potassium and include arrhythmias as well as neuromuscular dysfunction.
   ii. ECG changes include peaked T waves, widened QRS complex, and loss of P waves. Wide complex VT, VF, or asystole may follow if untreated.
   iii. Common causes in children:

      a. Hemolysis (pseudohyperkalemia – commonly seen with pediatric venipuncture)
      b. Excessive intake
      c. Renal impairment
      d. Succinylcholine

   iv. Treatment may be emergent in the face of arrhythmias.

      a. Calcium chloride (membrane stabilizer)
      b. Sodium polystyrene enema
      c. IV loop diuretics
      d. IV Na HCO 3
      e. Hemodialysis

8. Calcium (Ca):

   A. Most Ca is concentrated within bone matrix.
   B. Ca levels dependent on serum albumin concentration, as calcium is protein bound within the bloodstream.
   C. Direct measurement of serum ionized Ca is the most accurate means of determining available extracellular Ca.
   D. Hypocalcemia:

      i. Symptoms include perioral paresthesia, carpal pedal spasm, tetany, or seizures.
      ii. Chvostek sign: twitching of facial muscles in response to tapping over the path of the facial nerve (cranial nerve VII).
      iii. Trousseau sign: carpopedal spasm in response to an inflated sphygmomanometer cuff.
      iv. ECG changes include prolonged QT interval, peaked T waves, and heart block.
      v. Common causes in children include:

         a. Hypoalbuminemia
         b. TPN infusion
         c. Sepsis
         d. Pancreatitis

        e. Renal impairment
        f. Neonates are initially hypoparathyroid

    vi. Treatment requires calcium supplementation.

9. Magnesium (Mg)

   Magnesium levels are regulated by intestinal absorption, renal excretion, and bone deposition or mobilization.

   A. Hypomagnesemia

      a. In the setting of *low* urinary magnesium excretion:

         i. Starvation
         ii. Malabsorption
         iii. Diarrhea
         iv. Excess laxative use

      b. In the setting of high urinary magnesium levels:

         i. Renal deficiency (ATN)
         ii. Aldosteronism
         iii. Diuretics
         iv. Hypercalcemia
         v. Phosphate depletion

   B. Hypermagnesemia

      a. In the setting of *normal* renal function:

         i. Magnesium overload (poisoning)

      b. In the setting of *impaired* renal function:

         i. Hyperparathyroidism (hypercalcemia begets hypermagnesemia)
         ii. Hypothyroidism
         iii. Dialysis
         iv. Maalox (Mg(OH)2)
         v. Milk of magnesia

10. Phosphorus (P)

   Phosphorus, like potassium, is largely contained within the intracellular compartment. Similar to magnesium, serum levels are dependent on gastrointestinal absorption, renal excretion, and bone metabolism. Most total body P resides with the bone. Renal tubular resorption is under the physiological control of PTH.

   A. Hypophosphatemia

      a. In the setting of *low* urinary phosphate secretion:

         i. Glucose administration
         ii. Malabsorption

      iii. Diarrhea
      iv. Ingestion of aluminum- or magnesium-containing antacids
      v. Respiratory alkalosis

   b. In the setting of *high* urinary phosphate secretion:

      i. Glucose, amino acid, bicarbonate uric acid wasting, e.g., refeeding syndrome
      ii. Hyperparathyroidism
      iii. Androgen or estrogen administration
      iv. Thiazide diuretics

B. Hyperphosphatemia

   a. In the setting of *normal* renal function:

      i. Rhabdomyolysis
      ii. Malignant hyperthermia
      iii. Phosphorus poisoning
      iv. Hypoparathyroidism

   b. In the setting of *impaired* renal function:

      i. Chronic renal insufficiency
      ii. Trauma and surgery with increased cellular turnover

11. Common formulas:

   A. Water deficit (mL) = 600 × wt. (kg) × [1−(140/Na)].
   B. Plasma osmolality = 2 × Na + glucose (mg%)/18 + BUN (mg%)/2.8.
   C. FeNa = 100 × (urine Na × plasma Cr)/(urine Cr × plasma Na).

      i. FeNa <1 consistent with pre-renal condition.
      ii. FeNa >3 consistent with renal condition.

   D. Sodium deficit mL. 20% NaCl = wt. (kg) × 0.2 × (140−serum Na).

## Special Situations

A. Burns

Parkland formula

Deficit = 4 mL/kg/% total body surface burn, ½ given 1st 8 h after the time of burn, 2nd ½ in next 16 h. This formula underestimates fluid requirements in severe burns. Should follow with urine output and does not take into account maintenance fluid requirements.

B. Pyloric stenosis

Depending on degree of dehydration, D5 ½ NS @150–175 mL/kg/24 h until the patient voids. Then add 10–15 mEq of KCl/L depending on serum K level.

C. Postoperative

Maintenance plus additional 25–100% based upon the extent of the surgery.

D. Trauma

Follow ATLS protocols, bolus of 20 mL NS/kg till stable ×2 then blood.

## Study Questions

1. The initial approximate hourly fluid requirements for a healthy 16 kg child is:
   a. 58 mL/h
   b. 52 mL/h
   c. 56 mL/h
   d. 32 mL/h

   Answer: (b) 10 × 4 plus 6 × 2 equals 52 mL/h.

2. Causes of hypokalemia are:
   a. Diarrhea
   b. Excessive vomiting
   c. Diuretics
   d. All of the above

   Answer: (d) All of the above are associated with the development of hypokalemia.

3. The most common electrolyte abnormality in the postoperative patient is:
   a. Hypokalemia
   b. Hyperchloremia
   c. Hyponatremia
   d. Hypernatremia
   e. Hypophosphatemia

   Answer: (c) Hyponatremia is the most common postoperative electrolyte anomaly. Hyponatremia may occur when water is given to replace sodium-containing fluids. Antidiuretic hormone is also excreted in response to surgery and trauma.

4. The most appropriate replacement crystalloid solution for a pancreatic fistula is:
   a. Normal (0.9 %) saline solution
   b. D5W
   c. ½ Normal saline solution
   d. 5% Albumin
   e. Ringer’s lactate

Answer: (e) Pancreatic fluid is rich in bicarbonate (90 mEq/L). Only Ringer's lactate solution contains bicarbonate (28 mEq/L). Additionally, the pH of normal saline solution is 6.9.

5. ADH receptors are located within the:
   a. Glomerulus
   b. Proximal convoluted tubule
   c. Loop of Henle
   d. Distal convoluted tubule
   e. Collecting duct

   Answer: (e) ADH receptors are located within the collecting duct and are responsible for free water absorption.

## Further Reading

Holcomb and Ashcraft's Pediatric Surgery, 7th ed; Elsevier: 2020
Narins RG, Jones ER, Stom MC, et al. Diagnostic strategies in disorders of fluid, electrolyte and acid-base homeostasis. Am J Med. 1982;72:496–520.
Principles of Pediatric Surgery, 2nd ed; Mosby: 2003
Schwartz's Principles of Surgery, 11th ed; McGraw-Hill: 2019

# Pediatric Anesthesiology and Opioid Use

**Yohannes B. Getachew**

**Key Points**

1) Understanding the three phases of anesthetic care (more appropriately called “perioperative care”) and preoperative optimization is key for excellent clinical outcome.
2) Administration of an anesthetic creates temporary and reversible physiological changes to the body. Understanding these changes and monitoring and mitigating strategies are the foundation to a successful anesthetic care.
3) A particular focus on the risk of permanent brain effect from anesthetics administered in young patients, which is under investigation, is warranted.
4) Every attempt should be made to minimize the use of opioids in the management of postoperative pain in children. No one is an exception, but teenage patients are especially at high risk for abuse of pain medications.

The goals of anesthetic management are reversible loss of consciousness, anxiolysis, perioperative amnesia in different combinations, and, if needed, analgesia and muscle relaxation. Depending on the procedure, a combination of these goals is selected and the anesthetic plan formulated. Therefore, the type of anesthesia a patient receives varies from a mild sedation (we use a different term for sedation, “monitored anesthesia care” or MAC; be careful not to confuse with another “MAC” that is very commonly used when measuring the potency of volatile anesthetic agents, “minimum alveolar concentration”) to general anesthesia or, in appropriate cases in children, spinal anesthesia.

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## Essentials of Anesthetic Management of Pediatric Patients

1. Fundamentals of anesthetic care in children

   a. "Children are not small adults"—children go through rapid growth from birth to adulthood. Understanding these anatomic and physiologic changes and planning the anesthetic care accordingly are an essential part of a successful anesthetic management.

   b. Understanding the pathophysiologic changes due to the surgical diagnosis or preexisting medical condition and adapting the anesthetic plan to those changes are key to a high-quality clinical outcome (infection, mass or biochemical effect from a tumor, obstructive lesions, etc.).

   c. Preparing for the changes that occur due to the surgical intervention and adjusting the anesthetic plan are also critical. This includes mode of pain control, degree of blood loss, single lung ventilation, changes to vital organ perfusion, physical compression to the heart or lungs during surgery, and other changes to the cardiopulmonary system, etc.

   d. Communication between the surgical and anesthesia team is another key to a successful outcome. This includes details of the surgical intervention, what to expect from the surgical and anesthetic intervention, developments during procedure, change of surgical plan, etc.

2. An anesthetic course, which is also referred to as the perioperative period, has three phases—preoperative, intraoperative, and postoperative.

   a. Preoperative period—the start of this period depends on how one defines it, but for practical purposes, the moment a patient and family agree with a surgeon to have surgery at the facility and the process of booking the case is initiated can be considered a beginning of the preoperative period. It ends when the patient is taken to the procedure room, at which time, the intraoperative period starts. The term "procedure" includes surgical as well as nonsurgical activities (including percutaneous interventional and/or radiological procedures).

      Emphasis of the preoperative period is on preparing/optimizing patient for the procedure with excellent clinical outcome as the goal. This includes systemic review and addressing preexisting conditions—cardiac, pulmonary, CNS, renal, hepatic, metabolic including diabetes, etc.—correcting electrolyte and hematologic issues, treating infections (decisions on to proceed with procedure or not on a patient with URI are a very common problem), and checking if NPO status on day of surgery is followed (follow the American Society of Anesthesiologists (ASA) recommendation).

      For some of the above conditions, the earlier an intervention to optimize the patient is started, the better (e.g., as soon as decision is made to proceed with surgery), as some health conditions require time to correct. The decision on what laboratory tests need to be performed is also made during this period. The anesthesiologist and their team make decisions on the type of anesthetic and what type of monitor, in addition to the basic monitors, to use (urine output, use of invasive monitors, etc.) in the preoperative period. This is also the phase where the anesthetic plan, expectations, possible adverse issues, use of special monitors like arterial and central line, use of blood products, etc. are discussed with parents (and patient if he or she is old enough to participate). This might take place in the preoperative area on day of procedure or a few days to weeks ahead of time at a visit to a pre-surgery clinic.

   Well-established guidelines and procedures and well-trained nursing staff are critical to an effective preoperative process. They are mostly responsible for preparing patients, medication reconciliation, establishing rapport with family and the young patients, and settling the patient and family on day of surgery before and after the surgical and anesthesia teams arrive.

b. Intraoperative period—starts when patient is brought into the surgical/procedure suite where the procedure will take place and ends when the patient leaves the suite.

   A time-out before start of procedure represents the final assurance of patient's identity, surgical procedure to be performed, site of procedure, and the availability of equipment necessary for the stated procedure.

   The more the anesthetic and surgical teams communicate their intentions and expectations and work as a team, the better the outcome.

c. Postoperative period—starts when the patient leaves the procedure room and extends up to 48 h after the intraoperative period ends. Here, the recovery room nursing staff is introduced to the patient. It is of critical importance that both anesthetic and surgical information is adequately and precisely transferred to the PACU staff. This way the nurse knows what was done, what to expect, and what interventions are ordered to handle developments.

   The two most common anesthetic-related issues in PACU are nausea/vomiting and pain. Relatively less common are respiratory, cardiovascular, and other vital organ dysfunctions that may be related to the surgical and anesthetic intervention.

## Pharmacology of Anesthetic Medications

1. Anesthetic agents target the central nervous system (CNS) for their effects. The effect on the CNS and, sometimes, a direct effect on other organs cause the undesired effects which the anesthesiologist must mitigate—hypotension is due to depressed CNS as well as a direct effect of the drugs on the vascular system. Hypoventilation is due to depressed CNS which can be made worse by positioning and the surgical intervention. Reduced urine output can be the result of hypotension caused by the anesthetic agents and/or direct pressure on the kidneys or their blood supply, etc.
2. All the direct and intended effects of the anesthetic agents are reversible, i.e., when the drug is eliminated from the body, normal physiologic function resumes.

That means in addition to the patient regaining consciousness and memory, there will be reversal of the analgesic effects. The possibility of permanent brain effects in younger patients is discussed in the following section.

## Use of Anesthetics and Sedatives in Infants and Toddlers

Even though there is evidence from animal studies that exposure of the developing brain to anesthetics/sedatives may have effect on behavior, learning, and memory, the topic is under study and there is no clear evidence that these changes are due to the anesthetics. The FDA, IARS (International Anesthesia Research Society), AAP, American Pediatric Surgical Association, and other organizations have formed "Smart Tots," a multi-year collaborative effort. The latest information is maintained on the SmartTots website [1].

The best approach is to weigh the risks and benefits of the proposed operation, particularly in patients younger than 3 years old. This recommendation does not assume there is a definite cause and effect relationship between anesthetics and adverse CNS effect in children.

Two important factors are patient age and duration of surgery. The older the patient, the less the presumed risk. "Does the surgery need to be done?" and "Can the surgery be postponed to another time?" are some of the questions surgeons must consider when consulting with parents. The longer the operation, the longer the exposure to anesthetics.

Even though some parents are aware and will raise this topic with the surgeon in the clinic, most are not aware, and this important discussion doesn't take place. As a result, parents may face this issue in the preoperative area on day of surgery and find themselves conflicted on whether they made the right choice to have the surgery done on their child that day.

If the surgery can wait until the child is 3 years or older without increasing risk of complications, postponing is preferred. A study is underway on children younger than 2 years having more than 2 hr of general anesthesia randomized into two types of anesthetics. Follow-up is at 3 years of age, and the primary outcome is IQ. Results are projected to be ready by 2025 [2].

## Additional Notes

### *Use of Opioids in Pediatric Patients*

In addition to the respiratory and cardiovascular adverse effects, the use of opioid analgesics is associated with dependence/addiction in patients, and pediatric patients are not exceptions. Effort should be made to use all available non-opioid pain control options. This includes local infiltration of surgical sites, regional anesthesia, peripheral nerve block, non-opioid oral and parenteral analgesics, and, equally important, discussion of expectations of pain and its management with patient and parents when possible.

The approach to pain control should be considered as use of minimal narcotic medications and not total elimination. Some patients will have such intense pain that it warrants the use of potent opioid analgesics.

Discussion of postoperative pain should be started in the surgeon's clinic as well as at the pre-surgery visit with parents and patient, depending on patient's capacity. A clear pain management plan, as well as the parents/patient understanding and expectation, avoids misunderstanding and anxiety about the level of pain, how much pain tolerance is expected, the medications to use, and adverse effects from these medications. The possible use of opioids should be part of the conversation preoperatively as well as the adverse effects including the possibility of dependence on the drugs. Having a well-informed parent/patient is the goal.

### A) Use of Local Anesthetic Agents

For the intraoperative and immediate postoperative period, the use of local anesthetics in the form of regional anesthesia (caudal, epidural, and spinal analgesic/anesthetics) and peripheral nerve block or PNB (isolated nerve infiltration—femoral, sciatic, brachial, etc.) will give a dense and effective analgesia. Most of the time, a catheter is placed in the epidural space and run as an infusion that can stay up to 3 days. The same can be done for PNBs. For the rest it will be a one-time block performed pre-, intra-, or post-op. In these cases, depending on the type of local anesthetic used and type of block, the duration of effective pain control varies. Local anesthetics vary in duration from about 1 h for lidocaine up to several hours for the long-acting agents like bupivacaine, ropivacaine, and tetracaine. The type of block also influences the duration, mostly a function of blood supply to the region where the local agent was injected and how quickly the agent can be absorbed from the area. Spinal and epidural, being areas with significant blood supply, will last less than a PNB for the same local agent. Some PNBs last up to 12–16 h. Understanding the duration of a block is helpful for parent/patient expectation and planning when to start supplemental analgesia.

There are various PNBs that can be done for thoracic and abdominal surgeries, and a conversation between the surgeon and anesthesiologist is key for choosing the appropriate block for the type of surgery.

For some of the surgeries, an appropriate block/infiltration by the surgeon may be more practical. In those cases, it is important to keep in mind the anatomy of the nerve path and distribution and that all tissue layers must be infiltrated including skin and fascia for an effective block.

### B) After the Local Anesthetic Wears Off

The planned treatment modality after the local wears off should be started before the patient starts to feel pain to build adequate tissue level ahead of time. Waiting till patient starts to feel pain may mean dealing with severe pain, as sometimes the local wear is abrupt and the pain intense. These series of events will prompt unnecessary and unplanned use of opioid medications.

C) Narcotic Use

In many surgeries the use of narcotic medications is unavoidable. Familiarity with the medications and dosages by the ordering physician as well as the nurse following the patient is critical. Narcotics can be given in different modalities—IV PRN, IV parent/patient- or nurse-controlled PCA, or IM. The IM use has dwindled significantly since administration causes discomfort and has no added benefit over the IV use.

D) Monitoring

The other component to the use of opioid analgesics is monitoring in the inpatient setting. The respiratory and cardiovascular systems must be monitored while the patient is on narcotics for early detection of adverse effects. At the minimum there should be a pulse oximetry that detects both $O_2$ level and pulse.

For outpatients, adequate information must be delivered to caregivers concerning detection of adverse effects as well as use of the minimal opioid dose necessary to control pain.

## Study Questions

1) In which group of patients should the anesthetic risk of effect on behavior, learning, and memory be of particular concern?

   a. Operations performed under spinal anesthetic
   b. Patients with cardiac anomalies
   c. Patients younger than 3 years old
   d. Patients receiving opioid medication

   Answer to Question 1: (c.) Patients younger than 3 years old. There is no clear evidence that anesthetics cause changes in the developing brain; there is however evidence from animal studies. Further studies are underway to quantify the effect in humans. The best consensus that can be drawn at this time is to recognize that the risks should be especially considered for children who are under 3 years old. If the operation is needed, it should be performed, but the surgical team should ask, could it be safely delayed until the child is older?

2) Which of the following is not a strategy to reduce opioid use in children during the perioperative period?

   a. Use of local anesthetics for peripheral nerve blocks
   b. Preparing the child and parent for the level of pain they are to expect
   c. Use of multiple drug classes in combination to reduce pain
   d. Withholding analgesics until after the local anesthetic wears off

Answer to Question 2: (d.) Withholding analgesics until after the local anesthetic wears off. Opioid analgesics are powerful and effective medications to treat pain and when used judiciously and safely are an integral part of perioperative care for major operations. However, opioids carry dangerous side effects of respiratory and cardiovascular depression and can result in physical dependence when there is not a plan to wean therapy. One of the most important strategies in reducing opioid use is to have clear communication with child and parents before and after the operation to properly prepare them for the pain that can be expected as well as the strategies to treat it. Use of multimodality different classes of medication, such as acetaminophen, nonsteroidal anti-inflammatory medications, and local anesthetics in concert with opioids, will allow lower doses or no doses of opioids to be used. Preemptive use of local anesthetics for local infiltration, regional anesthetic, and local nerve block should be used to reduce the necessity for opioids.

## References

1. SmartTots, Funding Anesthesia Research To Ensure Pediatric Safety, 2021. https://smarttots.org/about/consensus-statement/. Accessed 20 Oct 2021.
2. SmartTots Panel on Pediatric Anesthesia Neurotoxicity Research. SmartTots. 2021. https://smarttots.org/smarttots-panel-on-pediatric-anesthesia-neurotoxicity-research/. Accessed 20 Oct 2021.

## *Further Reading*

Cravero JP, Agarwal R, Berde C, Birmingham P, Coté CJ, Galinkin J, Isaac L, Kost-Byerly S, Krodel D, Maxwell L, Voepel-Lewis T, Sethna N, Wilder R. The society for pediatric anesthesia recommendations for the use of opioids in children during the perioperative period—Cravero. Pediatr. Anes. 2019;20(6):547–61.

# Procedural Sedation

**Richard L. Lambert**

**Key Points**

1. A careful history and examination of the child is essential and should not be rushed. Inquiring about certain risk factors that can potentially complicate procedural sedation is important. These include previous history of difficulty with intubation or anesthesia, snoring, presence of rhinorrhea, or cough. The examiner should assess neck mobility, mouth opening, dentition, and the presence of facial or airway anomalies.
2. The class of medicines (opioids, benzodiazepines, ketamine, etc.) does not solely determine the depth of sedation or risk for complications. The dosing of these medications and the way they are administered or co-administered determine the safety and efficacy of the procedural sedation.
3. Any provider administering sedation must be able to adequately "rescue" a child from a complication or medication-related adverse effect. This includes the need to "rescue" a child that transitions into a deeper level of sedation than intended.

1. Procedural sedation
    (a) The use of procedural sedation outside the operating room to minimize or alleviate pain and/or anxiety has become commonplace in most children's hospitals.

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(b) Ideally, there should be separate providers assigned to the procedural sedation and to the procedure. This separation ensures that adequate attention can be given continuously to a sedated child. While moderate to deep sedation require the separate sedation provider, minimal sedation can often be safely achieved with a nurse monitoring the child and the surgical team performing the procedure. In the rest of the text, the person providing the procedure (surgeon) will be known as the proceduralist.

(c) The approach should be well thought out ahead of time, and the person providing sedation should be qualified to handle any situation that may arise during the sedation, including respiratory or cardiac decompensation, anaphylactic reaction, and vomiting with aspiration, to name a few.

(d) The process of providing procedural sedation can be divided into the following three components which will be discussed later in detail:

   (i) Pre-sedation
   (ii) Intra-procedure
   (iii) Post-sedation

2. Definitions and levels of sedation

(a) The approach to the child and use of medications may vary greatly, i.e., intramuscular (IM), intranasal (IN), or oral midazolam to relieve anxiety for a brief non-painful procedure versus intravenous fentanyl and propofol to achieve deep sedation for a procedure that is both painful and noxious.

(b) The term "conscious sedation," while still present in the medical vernacular, is a misnomer and should be replaced with "procedural sedation." A child is either conscious and receiving medications to relieve pain and/or anxiety, or he or she is sedated, with a correspondingly altered mental status (see Tables 1 and 2).

(c) Minimal sedation typically occurs after single doses of medications like oral, IN, or IM versed. Intranasal dexmedetomidine is commonly used for this purpose as well. Minimal sedation may also occur with a single small dose of opioid or ketamine. The goal of minimal sedation is anxiolysis or pain control without affecting the child's level of consciousness.

**Table 1** Common definitions used in procedural sedation

| | |
|---|---|
| Analgesic | Relieves pain by altering perception of nociceptive stimuli |
| Sedative | Decreases anxiety and calms patient |
| Anxiolytic | Relieves apprehension and fear due to anticipated act or illness |
| Amnestic | Affects memory; patient is unable to recall events after delivery of drug |
| Hypnotic | Produces drowsiness and aids in the onset and maintenance of sleep |

**Table 2** Levels of sedation

| | Minimal | Moderate | Deep | General anesthesia |
|---|---|---|---|---|
| Cognitive function | Normal to partially impaired | Partially to fully impaired | Fully impaired | Fully impaired |
| Responsiveness | Normal response to verbal commands | Purposeful response to verbal commands and/or light touch | Purposeful response to painful stimulation | No response to stimulation |
| Airway patency | Normal | May require intervention | Occasionally require intervention | Often require intervention |
| Spontaneous breathing | Normal | Normal to adequate | May be impaired | Often impaired |
| Cardiovascular function | Normal | Normal | May be impaired | Occasionally impaired |

(d) Moderate sedation usually requires IV access; however, a large dose of oral, IN, or IM sedative may lead to an altered mental status such that the child appears to be sleeping but opens eyes to voice and touch. Like with minimal sedation, these children rarely require airway manipulation, but should be monitored closely for cardiorespiratory changes and the potential to transition into deep sedation.

(e) Deep sedation requires IV access, both for drug administration and safety. Children in a state of deep sedation will not awaken, even to noxious stimuli. However, they should respond with purposeful movements during painful procedures, i.e., reach toward stimuli or move away from stimuli. Although uncommon, respiration may become impaired and bradycardia and/or hypotension can occur. A provider administering deep sedation should be qualified to maintain a pediatric airway, including BVM, oral/nasal airway, and potentially endotracheal intubation.

(f) General anesthesia is not administered routinely by procedural sedation teams outside of the operating room. This level of sedation is discussed in the chapter titled Pediatric Anesthesiology.

3. Pre-sedation assessment

   (a) History:

      (i) A general history of each child should be known prior to administering sedative and analgesic medications. Important historical information that may impact the choice of medications administered is summarized in Table 3.

      (ii) NPO guidelines per Table 4.

**Table 3** Pre-sedation assessment

| |
|---|
| Active medical issues |
| Current medications |
| Known allergies |
| Known adverse reactions to medications |
| Previous history of sedation or anesthesia and any complications |
| History of significant snoring or apnea |
| Known anatomic limitations to opening mouth or moving head and neck |
| Loose teeth or presence of dental devices |
| Time since last oral or enteral intake |
| Recent illnesses (fever, upper respiratory infection, etc.) |
| Family history of problems with sedation or anesthesia |

**Table 4** NPO guidelines

| Food | Hours of NPO |
|---|---|
| Clear liquids | 2 |
| Breast milk (infants) | 4 |
| Formula or light meal | 6 |
| Full meal | 8 |

(b) Physical exam:

  (i) A focused physical exam should include assessment of vital signs, facial and neck anatomy, cardiopulmonary function, and baseline neurological status.
  (ii) Airway (Fig. 1).

    a. A child’s tongue is relatively larger compared to the space it occupies in the oropharynx.
    b. Children with macroglossia, micrognathia, or retrognathia may have even more limited space relative to the oropharynx.
    c. Larynx is more anterior and superior.
    d. Relative to adult’s epiglottis, a child’s is longer, more narrow, and floppier.
    e. Children younger than 8 years old have the narrowest part of their upper airway located in the subglottic region at the level of the cricoid cartilage.
    f. Any obstruction in this area will significantly impair airflow.
    g. When supine, care should be taken to ensure the airway remains aligned and excess flexion or extension of the neck does not occur.

h. It is important to align the oral-laryngeal-pharyngeal axes. In the supine position, alignment is not optimal and upper airway obstruction may be present. A roll under the neck and/or shoulders should be utilized to create a “sniffing” position, whereby the horizontal plane of the ears (with patient lying supine) is anterior to

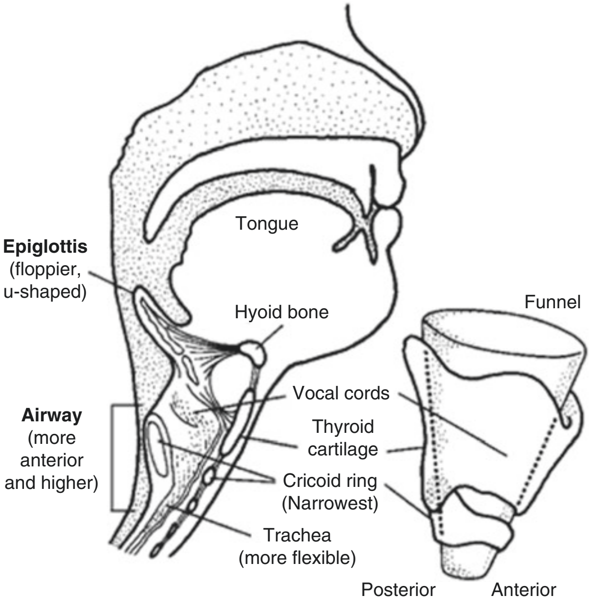
Tongue
Epiglottis
(floppier,
u-shaped)
Hyoid bone
Funnel
Vocal cords
Airway
(more
anterior
and higher)
Thyroid
cartilage
Cricoid ring
(Narrowest)
Trachea
(more flexible)
Posterior
Anterior

**Fig. 1** Pediatric airway, as compared to the adult. The narrowest part of the pediatric airway is the cricoid ring. (Included with permission from Susan Gilbert as published at http://www.medartist.com/emergency_pediatrics.html. Downloaded 8/17/21)

the shoulders. This position of the head and neck is "neutral position" (neither excessively flexed or extended). Infants have a large occiput and should have a roll placed under their neck and shoulders, whereas the older child will often require a roll under their head.

i. Mallampati classification may be performed to assist in predicting a difficult intubation but has not been shown to affect risk stratification for children undergoing procedural sedation. To perform the test, ask the child to sit up and open their mouth as wide as possible while sticking out their tongue. No vocalizations (don't ask the child to say "ah") should be performed as this changes the position of oral-pharyngeal structures. See Fig. 2.

(iii) In addition to the airway assessment, attention should be paid to any physical attributes the child may have that would put him or her at risk for complications, such as:

a. Limitation of neck movement
b. Obesity or failure to thrive
c. Baseline neurologic impairment
d. Scoliosis or other skeletal abnormalities

(iv) A brief examination of the chest with auscultation of the lungs and heart should be performed.

(c) After completion of the physical exam, assignment of an ASA score may be helpful in stratifying the child's relative risk of sedation (Table 5).

(d) Time-out should now occur. Ideally this will take place with parent(s) in the room. The purpose of this time-out is to verify:

  i. Patient identifiers confirmed.
  ii. Correct procedure is being done on the correct patient.
  iii. Consent has been signed (both for procedure and sedation).
  iv. Allergies have been reviewed.
  v. All members of the team should provide a verbal agreement to the time-out prior to proceeding with the procedure and sedation.

4. Intra-procedure

  a) Monitoring and equipment

    i. All children undergoing sedation to facilitate invasive or noninvasive procedures require continuous cardiopulmonary monitoring including pulse oximetry.

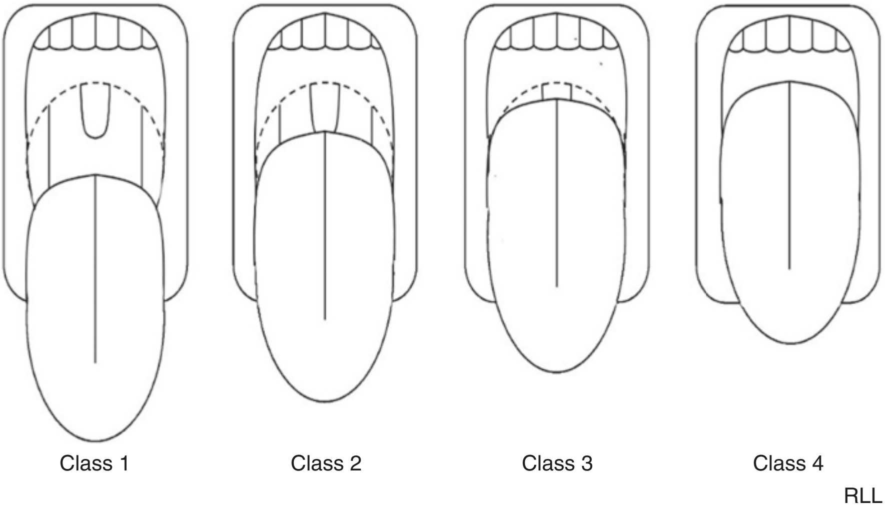

**Fig. 2** Mallampati classification. Class 1 reveals faucial pillars along with uvula and soft palate. Class 4 no longer has uvula or soft palate visible

**Table 5** ASA classification

| | |
|---|---|
| ASA I | Normal healthy patients |
| ASA II | Patients with mild systemic disease |
| ASA III | Patients with severe systemic disease that is limiting but not incapacitating |
| ASA IV | Patients with incapacitating disease which is a constant threat to life |
| ASA V | Moribund patients not expected to live more than 24 h |
| ASA VI | A declared brain-dead patient whose organs are being removed for donor purposes |

ii. Capnography to assess expired $CO_2$ is very useful in monitoring airway patency and ventilation in the sedated child.
iii. The mnemonic SOAPME can help the provider ensure they have the appropriate equipment in place prior to starting the procedure.
   a. S (suction)
   b. O (oxygen)
   c. A (airway)
   d. P (pharmacy)
   e. M (monitors)
   f. E (extra equipment)

b) Sedative medications (Table 6)

i. There are many sedatives that are currently used for procedural sedation. Some have purely sedative effects, while others also provide analgesia. When used in conjunction with an opioid or sedative, dosing should be decreased to minimize the chance of oversedation.
ii. Combinations include midazolam/fentanyl, midazolam/ketamine, propofol/fentanyl, and propofol/ketamine.
iii. Midazolam (Versed) is a benzodiazepine that has sedative, hypnotic, amnestic, and anxiolytic properties. It produces its effects by enhancing the inhibitory action of GABA.
   a. Commonly used as intramuscular (IM) injection for the child who either needs a brief procedure where complete stillness is not an issue or as premed to relieve anxiety for IV placement. Recommended max dosing is 10–15 mg total if given oral or IM.
   b. May also be given oral or nasal, but sneezing or coughing can limit its effectiveness and may require redosing.
   c. Flumazenil is a GABA receptor antagonist and can reverse the clinical effects of any benzodiazepine if overdose is suspected.

**Table 6** Sedative medications

| Drug | Dose | Onset (min) | Duration |
|---|---|---|---|
| Midazolam IV | 0.05–0.1 mg/kg | 0.5–1 | 30–60 min |
| Midazolam IM | 0.2–0.5 mg/kg | 5–15 | 30–120 min |
| Lorazepam IV | 0.03–0.1 mg/kg | 2–3 | 2–4 h |
| Ketamine IV | 0.5–2 mg/kg | 1–2 | 15–30 min |
| Ketamine IM | 2–5 mg/kg | 5–15 | 15–30 min |
| Propofol IV | 0.5–2 mg/kg | 0.5–1 | 5–10 min |
| Dexmedetomidine IV | 0.3–1 mcg/kg | 10–20 | 30–60 min |

iv. Lorazepam (Ativan) has similar clinical effects as midazolam.

  a. Rarely used for procedural sedation and cannot be given IM.
  b. Duration much longer.

v. Ketamine is a PCP derivative that has sedative, amnestic, and analgesic effects. It produces its effects by blocking NMDA receptors, leading to inhibition of glutamate-mediated transmission.

  a. Can be used as IM or IV injection alone to achieve both anxiolysis and analgesia, such as is needed for fracture reductions.
  b. May cause tachycardia and hypertension.
  c. May increase airway secretions.
  d. May cause airway bronchodilation and reduce risk of bronchospasm.
  e. Emergence reaction can occur as hallucinations, confusion, and/or agitation. Benzodiazepines in small doses may be given to help minimize this response.

vi. Propofol (Diprivan) is an anesthetic medication that has sedative, hypnotic, and amnestic effects. It is thought to produce its effect via the GABA pathways.

  a. Most commonly used sedative medication in procedural sedation.
  b. Typically used as bolus injections until induction occurs and then as IV infusion to maintain the desired level of sedation.

    (i) Infusion rates for procedural sedation range from 75 to 200 mcg/kg/min.
    (ii) Lidocaine (10 mg in 1 mL) can be left to dwell in PIV for 1 min before injection to alleviate local burning sensation. A tourniquet should be applied proximal to the IV site and left to remain tight around the extremity for 1 min. Following release of the tourniquet, the propofol induction can immediately begin. 10 mg of lidocaine should be included in the total weight-based dose allowed for the child during their procedure.

  c. Rapid onset of action and rapid recovery of consciousness after cessation of drug.
  d. May cause hypotension and/or apnea, especially with rapid bolus.
  e. Propofol infusion syndrome (PRIS) is a rare but serious adverse effect that comprises severe metabolic acidosis, hyperkalemia, rhabdomyolysis, and cardiovascular collapse and is frequently fatal. This effect has not been associated with brief procedures, rather, mostly in the setting of prolonged high-dose infusions.

vii. Dexmedetomidine (Precedex) is an extremely potent alpha 2 receptor agonist with preference for centrally mediated increase in GABA activity. It produces sedation and analgesia.

**Table 7** Analgesic medications

| Drug | Dose | Onset (min) | Duration |
|---|---|---|---|
| Morphine IV | 0.05–0.1 mg/kg | 2–5 | 1–2 h |
| Morphine IM | 0.1–0.2 mg/kg | 5–10 | 1–2 h |
| Fentanyl | 0.5–1 mcg/kg | 0.5–1 | 15–30 min |
| Hydromorphone | 0.01 mg/kg | 10–15 | 4–6 h |

a. May be given as a bolus followed by an infusion, or without a bolus given its onset of activity is very rapid.
b. Boluses should be given very slowly and not more frequent than every 10 min.
c. Rapid bolus can lead to dramatic bradycardia and hypotension.
d. Bradycardia is common during boluses and longer duration of infusion.

c) Analgesic medications (Table 7)

i. Most analgesics used for procedural sedation are opioid based. Non-opioid-based analgesics such as nonsteroidal anti-inflammatory drugs (NSAIDs) and acetaminophen are typically reserved for use after the procedure. However, Toradol, an intravenous NSAID, when administered prior to the start of sedation for a painful procedure, may decrease need for opioids both during and after procedure.
ii. If an adverse effect such as severe respiratory depression is suspected in a child who received an opioid, naloxone should be given. This will reverse the analgesic and sedative effect for a brief period. Be prepared for the child to awaken and be in obvious pain. Also be prepared to redose as necessary until the symptoms of overdose have resolved.
iii. Morphine is the prototypic opiate and acts on the mu receptor to induce analgesia. Higher doses will also cause sedative and hypnotic effects. It can be given PO, PR, SQ, IM, or IV. Dosing varies by route and should be verified prior to administration.

    a. High doses in infants have been associated with prolonged apnea.
    b. Histamine release may lead to local flushing and pruritus as well as systemic effects like wheezing or hypotension.

iv. Fentanyl is a synthetic derivative of piperidine that acts at the mu receptor. It has less hypnotic and sedative effects than morphine.

    a. As much as 100 times more potent than morphine.
    b. Has a very rapid onset.
    c. Should be bolused very slowly (over 2–5 min) to minimize the chance of chest wall rigidity and respiratory decompensation.
    d. May be used in the child with suspected allergy to morphine.

        v. Hydromorphone (Dilaudid) is a derivative of morphine that acts at the mu receptor.
            a. As much as five times more potent than morphine.
            b. Has the longest duration of the opioids.
            c. Like fentanyl, causes less pruritus, nausea, and dysphoria than morphine.
    d) Intraprocedure communication
        i. Defined expectations should be discussed between team members prior to the start of the sedated procedure. This will markedly decrease the potential for unplanned events for both the proceduralist and the sedation provider.
        ii. The proceduralist should communicate his or her plan to inject local anesthetic (if applicable) so that the sedation provider may be prepared for the possibility of needing to give a bolus of sedative to prevent excessive movement. Likewise, it is helpful to know when any other painful procedures commence. Boluses of analgesic may be appropriate at this time.
        iii. Finally, a "heads up" regarding estimated time left in the procedure can be very helpful to the sedation provider. They can use this information to begin decreasing infusion rates, determine if new syringes need to be prepped, or augment the ongoing sedation regimen to prepare for awakening the child.

5. Post-sedation
    a) The child should be monitored until he or she returns to his or her pre-sedation baseline.
    b) A child should not be discharged until certain criteria are met:
        i. Stable vital signs.
        ii. Return to baseline mental status.
        iii. Head control and strength are appropriate to maintain a patent airway.
        iv. Pain is well controlled.
        v. Nausea is well controlled.
    c) Special circumstances such as administering a reversal agent or a prolonged adverse event during the sedation may require the child to be monitored longer and/or admitted to the hospital for overnight observation.
    d) Specific instructions should be given to the child's family telling them what to do if the child has any medical problems immediately following the sedation.

6. Adverse events during procedural sedation
    - a) Desaturation may occur from many causes and should be treated immediately. The most common cause is secondary to upper airway obstruction. However, lower airway obstruction, especially in the child with known asthma, may occur suddenly and require treatment.
        - i. Check equipment to ensure oxygen is flowing and all tubes are connected and without obstruction.
        - ii. Apnea can be central or obstructive.
            - a. Central apnea is usually a result of oversedation but may occur even with standard doses.
                - (i) Treatment consists of supporting the child's airway and breathing until the medication effect has worn off.
                - (ii) May require bag mask ventilation.
                - (iii) May consider reversal agent if prolonged.
            - b. Obstructive apnea is obvious by the observation that child has abdominal and/or chest movements but no air entry into the lungs.
                - (i) Treatment is based on the specific causes that will be discussed below.
                - (ii) Upper airway obstruction (UAO) is the most common cause of desaturation and is usually due to hypotonia of soft palate and/or epiglottis, caudal displacement of the tongue, accumulation of secretions, or laryngospasm. There are two types of sounds that can be heard during UAO: stertor and stridor.
                    - a. Stertor is a low-pitched sound that originates most commonly from the pharynx. Stertor is a Latin derivation of "to snore," and its characteristics match that well-known sound. It may occur with inspiration, expiration, or both.
                    - b. Stertor should be addressed immediately by suctioning of the oropharynx and possibly nasopharynx, using caution not to suction too deep and cause coughing or reflex gagging. Simultaneous repositioning of the head and neck should occur as well.
                    - c. If pharyngeal hypotonia is suspected, reposition the head and neck to ensure midline position with slight extension of neck: Do not overextend.
                    - d. "Sniffing position" as discussed earlier in the chapter is the desired position to maintain a patent airway.
                    - e. If midline position is not successful, gently rotating child's head to the side while maintaining slight extension of neck may be tried.

  f. Provide chin lift followed by jaw thrust until obstruction has been relieved.
  g. Consider nasal trumpet or oral airway as adjuncts.

iii. Laryngospasm is a spasm of the glottic structures and can be an emergency if complete; partial obstruction allows some air entry. The noise that occurs during a laryngospasm is more consistent with stridor. Unlike stertor, stridor is a high-pitched sound caused by severe narrowing of the larynx, either from inflammation, swelling, or laryngospasm. This sound typically occurs with inspiration but can occur during expiration or both. Severe laryngospasm may occur with little to no audible sounds.

  a. May occur during induction or emergence but can occur at any time.
  b. Stridor may be heard and is often pre-empted by coughing.
  c. If complete, child will be attempting to breath, but no air entry is occurring.
  d. Prolonged attempts to breath with sudden release of glottic obstruction can cause pulmonary edema.

iv. Treatment is multifactorial and stepwise, including:

  a. Increase in oxygen supply immediately.
  b. Jaw thrust with suctioning of the mouth and oropharynx.
  c. Deepen sedation with bolus of IV sedative.
  d. Pressure on the “laryngospasm notch,” an area just anterior to the mastoid process and below the earlobe, can be effective while performing a jaw thrust. It is important that pressure not be applied on the mandible, but rather the most cephalad portion of the laryngospasm notch. Done properly, this maneuver should create brief but severe pain from direct pressure on/near the facial nerve and parotid gland. If done correctly, and in conjunction with a proper jaw thrust, the laryngospasm typically transitions into stridor in one to two breaths followed by unobstructed breathing in another one to two breaths.
  e. If none of the above are successful within 15–30 s, bag mask ventilation must be applied starting with continuous positive airway pressure (CPAP) and administering positive pressure breaths if necessary.
  f. Finally, if the child remains without adequate chest expansion and vital signs, consider a short-acting muscle relaxant such as succinylcholine. This will allow you to use BVM and attain chest rise and gas exchange, but patient will no longer be maintaining his or her airway, and nor will he or she have spontaneous respiratory effort until paralytic effect resolves.

b) Aspiration of oropharyngeal or gastric contents can occur at any time during the procedural sedation.

  i. Proactive suctioning of secretions immediately upon hearing airway noises may be preventative.
  ii. Minor aspiration will not require treatment other than the patient clearing his or her airway with a productive cough.
  iii. A major aspiration, especially with evident vomitus, will likely require abortion of the procedure with subsequent treatment including:

    a) Increased oxygen supply including bag-valve-mask.
    b) Albuterol treatment may help with airway clearance.
    c) Monitoring after a child has awakened may include hospital admission and observation for signs and symptoms of aspiration pneumonia or pneumonitis.

c) Hypotension may occur as a direct result of the sedative medications, as a secondary result of the child's underlying medical condition, or a combination of both. Hypotension may be a result of peripheral vasodilatation, direct myocardial depression, or both. Changes in cardiac output from changes in intra-thoracic pressure during sedation may also occur.

  i. A drop in BP from baseline is extremely common when using propofol and will be exacerbated by any adjunct medications such as an opioid. A drop of 10–15% from baseline is well tolerated and generally does not require interventions.
  ii. If BP continues to drop, consider a decrease in the sedative infusion rate and a fluid bolus, i.e., 10–20 mL/kg NSS.
  iii. Reassessment of the child, including palpation of pulses and examination of skin, should occur to ensure perfusion is maintained.

d) Bradycardia may occur during moderate to deep sedation. Like the drop in BP, a decrease of more than 15% from baseline should warrant further investigation including evaluation of the rhythm for any abnormal beats or changes in QRS complex.
e) Anaphylaxis or allergic reaction is rare but can be life-threatening. Any existing allergies should be identified during the pre-assessment phase.

  i. Urticaria with or without redness that was not present prior to the procedure along with any signs or symptoms of respiratory distress and swelling of the face and/or tongue should be considered an allergic reaction.
  ii. Treatment consists of the following:

    a) Abort the procedure and administer 100% oxygen.
    b) Stop administering the sedative/analgesic.
    c) Administer epinephrine 0.01 mg/kg of 1:1000 concentration IM into the lateral thigh; may need to repeat.
    d) Give 10–20 mL/kg NSS bolus, may repeat if necessary.

e) Consider antihistamines (H1 and H2 blockers).
f) Consider IV corticosteroids.
g) If not responding immediately, get help and provide cardiopulmonary support until help arrives.

## Additional Notes

Procedural sedation for children can be done safely and with great success. The key is to be thoroughly prepared for any eventuality. Know your equipment, know your medications, and know the personnel with whom you are working. Have a plan prior to starting the procedure, and communicate regularly to the sedation provider during the procedure. Finally, knowledge about your patient and his or her medical history is a must. Remember, safety first, before, during, and after the procedure!

## Study Questions

1) Question 1: Anxiolytics are defined as those which:

   a. Produce drowsiness and aid in the onset and maintenance of sleep.
   b. Relieve pain by altering perception of nociceptive stimuli.
   c. Affect memory; patient is unable to recall events after delivery of drug.
   d. Relieve apprehension and fear due to anticipated act or illness.

   Answer to question 1: (d.) Relieve apprehension and fear due to anticipated act or illness. Many medications used for sedation can provide anxiolysis through different mechanisms. Some, such as opioids, at high enough doses, will alter a child's mental status such that he or she becomes tired or distracted from the stimulation at hand. However, the main intent of using an opioid is for pain control. Other drugs such as ketamine, benzodiazepines, or propofol can elicit hypnotic, amnestic, and sedative properties as well. The goal of providing anxiolysis involves relieving apprehension and fear associated with the procedure. Anxiolysis is not typically considered a level of sedation but can sometimes affect a child in a manner that would be consistent with "minimal sedation."

2) Question 2: Which of the following levels of sedation is characterized by an inability to awaken to voice, presence of a purposeful response to painful stimulation, and a potential for impaired respiration?

   a. Deep sedation
   b. Moderate sedation
   c. General anesthesia
   d. Minimal sedation

Answer to question 2: (a.) Deep sedation. The concept of structured levels of sedation is based on a continuum of altered mental status that ranges from mild impairment and no hemodynamic effects while under minimal sedation to complete loss of consciousness, no airway protection, and high potential for hemodynamic effects achieved under general anesthesia. It is common to have a child under one level of sedation suddenly become more deeply sedated, changing his or her risk for adverse events and forcing the provider to "rescue" him or her from the unintended deeper level of sedation. The goal of deep sedation is to achieve a state whereby a child is not awake but can move appropriately to noxious stimuli and is able to maintain his or her airway without external support. It should be noted that interventions, such as suctioning, repositioning, or even jaw thrust, to open an upper airway in a child undergoing deep sedation are not uncommon.

3) Question 3: Which of the following terms best describes the ideal positioning of the head and neck to align the upper airway during sedation?

   a. Laryngospasm maneuver
   b. 3:3:2 rule
   c. Sniffing position
   d. Neutral alignment

Answer to question 3: (c.) Sniffing position. A laryngospasm maneuver is a procedure whereby a provider uses his or her fingers to apply firm pressure at the "laryngospasm notch" located anterior to the mastoid process and just below the earlobe. This maneuver creates severe pain and is thought to lead to relaxation of the vocal cords via vagal nerve stimulation and autonomic modulation. Pressure is applied at the same time the provider is pushing the mandible forward in a jaw thrust maneuver. The 3:3:2 rule is part of an evaluation of a patient's neck anatomy used for predicting a difficult airway. The first 3 refers to the opening diameter of a patient's mouth (placement of three fingers between incisors). The second 3 is also determined using three fingers and is achieved by measuring the distance between the anterior tip of the mandible and the neck of the hyoid bone. The 2 refers to placing two fingers between the floor of the mandible and the thyroid notch. Difficult airways are typically associated with failure to achieve the 3:3:2 designation. This rule does not apply to smaller children. A neutral head position occurs when the neck is straight, neither flexed nor extended, and in alignment with the rest of the body. The sniffing position utilizes the neutral head position except there is additional positioning of the head in an anterior direction so that the ears are located anterior to the front of the shoulders. This position best aligns the oral-laryngeal-pharyngeal axes to achieve an open upper airway.

4) Treatment of laryngospasm is multifactorial and stepwise. Put the following steps in correct order. 1. BVM applied to the fact to provide CPAP. 2. Increase oxygen. 3. Pressure on the “laryngospasm notch.” 4. Jaw thrust with suctioning of the mouth and oropharynx.

   a. 2,4,3,1
   b. 4,3,2,1
   c. 1,2,3,4
   d. 3,2,1,4

   Answer to question 4: (a.) 2, 4, 3, 1. Laryngospasms may occur at any time during a sedation. It is most common during induction with deep sedation but can also occur when inducing or recovering from general anesthesia. If the obstruction is complete, the patient will not be able to inspire, and respiratory failure can occur. Treatment must be immediate and is most successful when done in a stepwise fashion. Supplemental oxygen should be the first intervention. Immediately following the application of oxygen, provider should simultaneously attempt suctioning of the mouth and oropharynx and perform a jaw thrust. With hands in place for the jaw thrust, a concurrent laryngospasm maneuver should be attempted. Finally, if this is not successful in providing adequate oxygenation, a mask should be applied to the child’s face with CPAP and/or PPV being administered until the child is able to breathe on his or her own. Many providers will also give a bolus of sedation at or near the onset of the laryngospasm.

5) The mnemonic SOAPME can assist a provider in ensuring he or she is adequately prepared to start a sedated procedure. Which of the following terms is not included in this mnemonic?

   a. Oxygen
   b. Action
   c. Extra equipment
   d. Pharmacy

   Answer to question 5: (b.) Action. There is no single thing more important to the success and safety of a sedated procedure in children than being adequately prepared for all potential outcomes. The SOAPME mnemonic is designed to help the provider remember several key aspects of this preparation and includes suction, oxygen, airway, pharmacy, monitors, and extra equipment. While it is necessary to understand potential actions that will be required during a sedated procedure, action is not part of the mnemonic.

## Further Reading

American Society of Anesthesiologists Task Force on Sedation and Analgesia by Non-Anesthesiologists. Practice guidelines for sedation and analgesia by non-anesthesiologists. Anesthesiology. 2002;96(4):1004–17.

Barbi E, et al. Deep sedation with propofol by non-anesthesiologists. Arch Pediatr Adolesc Med. 2003;157:1097–103.

Berde CB, Sethna NF. Analgesics for the treatment of pain in children. N Engl J Med. 2002;347:1094–103.

Coté CJ, Wilson S. Guidelines for monitoring and management of pediatric patients before, during, and after sedation for diagnostic and therapeutic procedures. Pediatrics. 2019;143(6):e20191000.

Cravero JP, et al. Incidence and nature of adverse events during pediatric sedation/anesthesia for procedures outside the operating room: report from the pediatric sedation research consortium. Pediatrics. 2006;118(3):1087–96.

Devlin JW, Roberts RJ. Pharmacology of commonly used analgesics and sedatives in the ICU: benzodiazepines, propofol, and opioids. Crit Care Clin. 2009;25(3):431–49.

Iyer MS, et al. Higher Mallampati scores are not associated with more adverse events during pediatric procedural sedation and analgesia. West J Emerg Med. 2018;19(2):430–6.

McMorrow SP, Abramo TJ. Dexmedetomidine sedation: uses in pediatric procedural sedation outside the operating room. Pediatr Emerg Care. 2012;28(3):292–6.

Tobias JD. Sedation and analgesia in paediatric intensive care units: a guide to drug selection and use. Paediatr Drugs. 1999;1(2):109–26.

# Postoperative Neonatal and Pediatric Critical Care

**Edward A. Everett Jr and Frank A. Maffei**

**Key Points**

1. Among the most essential tenets of pediatrics is the understanding of the physiologic and anatomic differences between the neonate, infant, child, and adult which provides a foundation for providing optimal postoperative ICU care.
2. The application of this broad understanding of the physiologic and anatomic differences between these different pediatric patient populations enables a safe and efficient transition of care from the operative/anesthetic setting to the ICU and guides ongoing monitoring and focused expectations and prognostication.

1. Outline
    (a) Developmental considerations
    (b) Transition of care from the operative/anesthetic setting to the ICU
    (c) ICU monitoring of the postoperative patient
    (d) ICU discharge criteria and transition of care to the ward and home

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2. Developmental considerations

   Among the most essential tenets of pediatrics is the understanding of the developmental differences that separate the neonate, infant, child, and adult. These differences are especially pronounced in the neonate and infant. A brief overview of important physiologic and anatomic differences between the neonate, infant, child, and adult will provide a foundation for providing optimal postoperative ICU care.

   (a) Airway

      (i) There are important anatomic and functional differences between the airway of a small infant and older child. Several characteristics of the neonate and infant airway increase the risk for upper airway obstruction.

         1. A prominent occiput causes flexion of a relatively short neck when lying supine.
         2. The tongue is proportionally larger.
         3. The epiglottis and soft tissues of the upper airway are more compliant.
         4. The larynx is more anterior and cephalad (C3–4 in an infant vs. C4–5 in an adult).
         5. Historically the larynx has been thought to be cone shaped and does not assume its cylindrical shape until approximately 8 years of age. However, recent data in neonates using MRI and bronchoscopy reveal a cylindrical larynx with an elliptical cross section and a slightly increased anteroposterior dimension. In some circumstances, if large enough, a round endotracheal tube can place undue pressure on the lateral mucosa.
         6. The neck is shorter and concomitantly the vocal cords lie at C4 in the neonates and infants while at C5 or C6 in older children and adults.
         7. Incomplete development of the cartilaginous rings throughout the conducting airways increases the potential for airway obstruction along the entirety of the airway.
         8. Mucosal edema of the airway is poorly tolerated. The narrowest part of the infant airway is at the cricoid ring in contrast to the vocal cord aperture in older children. Poiseuille's law states that resistance to air flow is inversely proportional to the fourth power of the radius of the airway; consequently, 1 mm of concentric edema at the level of the cricoid ring increases resistance 16 times.
         9. Laryngomalacia is a common finding in preterm infants.
         10. Infants are obligate nose breathers. Nasal passages may become completely or partially obstructed with mucous, edema, or a large nasogastric tube.

(b) Breathing

  (i) The diagnosis and management of postoperative respiratory insufficiency in small infants and neonates require a clear understanding of the developmental anatomy and physiology of the thorax and lungs. In addition, an immature central nervous system also can often influence control of breathing.

    1. Maintenance of functional residual capacity (FRC) is of vital importance considering that FRC is very close to closing volume (CV) in infants. This is especially true in preterm infants. FRC is the volume of gas within the lung at the end of tidal breathing. It is achieved when the inward forces of the lung balance the outward forces of the chest wall. CV is the volume above residual volume at which the small airways and alveoli begin to collapse. Because CV is very close to FRC, infants have an inherent tendency to have alveolar collapse (atelectasis). This propensity for atelectasis is especially notable during sedation and anesthesia and in preterm infants at risk for central apnea.
    2. Challenges to maintaining functional residual capacity (FRC) in the small infant.

        (a) The chest wall is far more compliant due to a cartilaginous rib cage and poor muscular development both decreasing outward elastic recoil.
        (b) Diminished compliance of the lung due to incomplete conducting airway and alveolar development combined with greater interstitial tissue matrix serve to increase inward elastic recoil. This is further accentuated in preterm infants by overt or residual surfactant deficiency. Diminished surfactant quantity or function increases air surface tension which can lead to end expiratory alveolar and terminal airway collapse.
        (c) Reduced density of type I muscle fibers when compared to older children and significant respiratory muscle inefficiency at high ventilatory rates increase the risk of early respiratory fatigue.
        (d) Unlike the adult, infants and neonates lack the large cross-sectional area of the distal airways, and thus, a larger portion of total airway resistance occurs in the peripheral small airways. Additionally, branching morphogenesis within the lung as well as alveolar septation can be permanently altered in preterm infants with severe bronchopulmonary dysplasia.

    3. Fortunately, there are several effective compensatory mechanisms during normal tidal breathing that act to maintain the infant's FRC.

        (a) Baseline increased minute ventilation allows short expiratory times resulting in intrinsic positive end expiratory pressure (PEEP).

- (b) Laryngeal braking is early glottic closure ceasing expiration prior to reaching closing volume, preventing small airway collapse. An exaggeration of the laryngeal braking phenomenon occurs during states of decreased compliance and is clinically manifested by end expiratory grunting.
- (c) Respiratory muscle and diaphragmatic tonicity during exhalation.

(c) Cardiovascular

- (i) Key events in the transition from the intrauterine to extrauterine cardiovascular function are summarized below:
  1. Expansion and oxygenation of the newborn lung causes a gradual reduction in pulmonary vascular resistance (PVR) in the first days after birth. However, PVR does not fall to adult levels until 2–4 months after birth.
  2. This drop in PVR together with a simultaneous increase in systemic vascular resistance causes the flap of the foramen ovale to close against the secundum atrial septum. Concomitantly, rising oxygen tension initiates closure of the ductus arteriosus. The complete anatomic closure of the ductus arteriosus occurs in 98% of infants by day 4 of age. Persistent patency of the ductus arteriosus, and to a lesser extent the foramen ovale, can lead to clinically significant intracardiac shunting.
  3. The neonatal myocardium is composed of poorly organized myocytes that have approximately half the amount of contractile elements (actin, myosin, troponin, and tropomyosin) that are present in older children. Neonates also have a functionally immature sarcoplasmic reticulum that leads to inefficient calcium transport into the myofibrils. Due to this immature myocardium, the infant heart has greater reliance on extracellular calcium. This explains the greater inotropic response to exogenous calcium and digitalis seen in newborns when compared to adults.
  4. Autonomic innervation of the myocardium is also functionally immature. Although the density of beta-adrenergic receptors is high, beta—receptor—adenylase cyclase coupling mechanisms are inefficient. Vagal innervation of the myocardium is complete at birth leading to a state of parasympathetic dominance. This is consistent with the neonate's propensity for bradycardia during stress.
  5. The ongoing development of the myocardium results in an infant with a poorly compliant ventricle that has limited responsiveness to increasing preload. The relative inability to increase stroke volume to increase cardiac output leads to a state of chronotropic dependence. Fortunately, infants are generally tolerant of tachycardia during periods of increased metabolic demand. This dependence on heart rate to maintain cardiac output is especially noteworthy when considering the infant's vulnerability to bradycardia secondary to central apnea of

prematurity or vagal stimuli. Despite the ongoing development of the cardiovascular system, the infant maintains an impressive baseline cardiac output to meet an already high metabolic demand. Although the absolute cardiac output as measured in L/min is low as compared to an adult, the cardiac output in relation to body size is higher in the neonate, measuring up to 4 L/min/$m^2$ as compared to 2.5–3.5 L/min/$m^2$ in the adult.

(d) Central nervous system

  (i) The infant brain is in a rapid phase of cellular development. Although infants possess a similar number of neuronal cells as adults, neurotransmission remains functionally immature due to ongoing axonal and dendritic growth. Glial cells (astrocytes, oligodendrocytes) continue to multiply well into the second year of life; they serve as neuronal support and are important in myelination. Myelination of axons is an ongoing process that begins early in prenatal life and extends to 6 years of age. As myelination advances in a rostrocaudal fashion, axonal impulse transmission becomes more efficient. In preterm infants the autoregulation of cerebral blood flow is poorly established especially early after birth. This represents a primary factor that predisposes the preterm infant to develop germinal matrix and/or intraventricular hemorrhage. Additionally, due to a watershed circulation directly impacting the periventricular white matter, hypoperfusion can lead to the development of periventricular leukomalacia. Both these adverse events carry potentially significant implications for future neurodevelopment. A major distinction from a gross anatomical basis is the presence of open fontanelles in the small infant.

  (ii) The anterior fontanelle remains open until 12–18 months, whereas the posterior fontanelle closes by 4 months. A common misconception is that due to the presence of open fontanelles, infants are fully protected from increases in intracranial pressures. This may in part be true in cases of slowly evolving intracranial hypertension (i.e., as may occur in chronic hydrocephalus), but not true during acute increases in intracranial pressure (i.e., as may occur during meningitis or in traumatic brain injury). The dura mater is poorly compliant and does not accommodate acute increases in intracranial pressures; therefore, the infant remains at risk for herniation syndromes during states of increased intracranial pressure.

  (iii) Central respiratory control of breathing is diminished under anesthesia and is associated with a markedly decreased ventilatory response to hypercarbia. In preterm infants, the potentially delayed clearance of anesthetic drugs combined with the maintenance of normal to low normal carbon dioxide levels while on mechanical ventilation perioperatively can exaggerate periodic breathing or frank apnea in the postoperative period (Fig. 1).

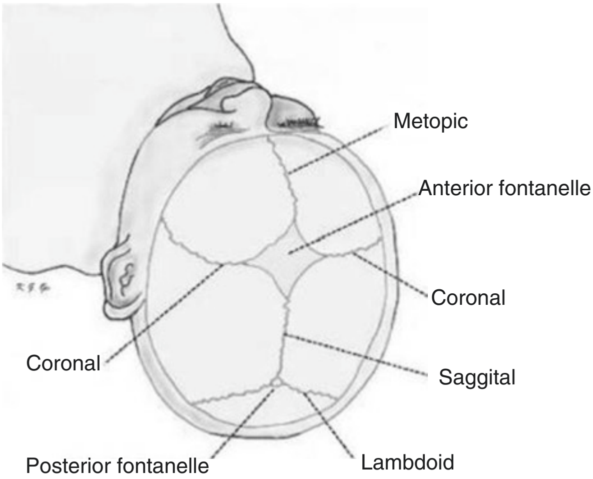

**Fig. 1** Cranial sutures and fontanelles. The soft spots between the cranial sutures are the anterior and posterior fontanelles. The fontanelles close by age 18 months old (*Source*: Adapted from xxjamesxx under Creative Commons Attribution-Share Alike Unported license as published on https://upload.wikimedia.org/wikipedia/commons/5/5f/Sutures_from_top.png. Downloaded 1 Jan 2014)

(e) Temperature control

(i) Infants and newborns have a large surface area to volume ratio that allows for greater heat loss. They also lack significant amounts of insulating body fat. If proper precautions are not taken, small infants are at profound risk for heat loss from conduction, convection, evaporation, and radiation. Important thermoregulatory mechanisms are immature and inefficient in the preterm and small infant. For example, brown fat which is produced during the third trimester is vital for thermogenesis. This lack of brown fat in small for gestational age and infants born preterm impedes the infant's innate ability to mount an adequate response to cold stress. In addition, small infants have a limited or absent ability to shiver. Since infants have immature thermoregulation, close attention to maintaining euthermia should be an essential component in the care of the postoperative patient. An important strategy might include policies which directly address the appropriate room temperature for pediatric and neonatal patients within the operative suit. Additionally, the development of protocols for inter-unit transport to mitigate risks of inadvertent hypothermia has also shown promise.

3. Transition of care from the operative/anesthetic setting to the neonatal or pediatric ICU

(a) The Joint Commission estimates that 80% of serious medical errors involve miscommunication during care transitions. Miscommunication was the leading root cause of sentinel events reported to the commission between 1995 and 2006. The majority of avoidable adverse events are due to the lack of effective communication. Recent data suggests that standardization of the postoperative handoff process of children undergoing surgery improves outcomes.

(b) Using the acronym SHARE, transitions can become more efficient, informative, and safe.

  (i) Standardize critical content, including:

    1. Providing details of the infant's history to the receiver
    2. Emphasizing key information about the infant when speaking with the receiver
    3. Synthesizing clinical information from separate sources before passing it on to the receiver

  (ii) Hardwire within your system, including:

    1. Preparing and stating expectations about how to conduct a successful handoff
    2. Developing standardized forms, tools, and methods, such as checklists
    3. Using a quiet workspace or setting that is conducive to sharing information about a patient

  (iii) Allow opportunities to ask questions, including:

    1. Using critical thinking skills and scrutinizing and questioning the data
    2. Sharing and *receiving information as an interdisciplinary team*
    3. Exchanging contact information in the event there are any additional questions

  (iv) Reinforce quality and measurement, including:

    1. Demonstrating leadership commitment to successful handoffs
    2. *Monitoring compliance* with use of standardized forms, tools, and methods for handoffs
    3. Using data to determine a systematic approach for improvement

  (v) Educate, including:

    1. *Teaching* staff and standardizing training on how to conduct a handoff
    2. Providing real-time feedback to staff

(c) Patient and data handoff

  (i) Patient is transferred from OR to ICU by following personnel:

    1. Attending anesthesiologist and or CRNA
    2. Primary surgeon
    3. Respiratory therapy

  (ii) Anesthesia to ICU staff report should include:

    1. Nature of airway and how intubation was performed
    2. Anesthetic course

        3. Intraoperative pulmonary function and current respiratory support
        4. Cardiovascular function, rhythm, and current hemodynamic support
        5. Intraoperative volume and/or blood product administration
        6. Last sedative/opioid/neuromuscular blocker

    (iii) Surgeon to ICU staff report should include:

        1. Review of preoperative lesion
        2. Intraoperative findings
        3. Description of repair
        4. Hemostasis
        5. Expected postoperative course including but not limited to:
            (a) Mechanical ventilation strategy (extubation versus continued support)
            (b) Need for further volume administration
            (c) Need for further blood product administration
            (d) Timing of wound closure, when applicable
            (e) Possible returns to OR (i.e., washouts, closures)
            (f) Specific organ system concerns
            (g) Perioperative antimicrobial regiment
        6. Current vascular access and drainage tubes
        7. Current medications
        8. Expected lab schedule

4. ICU monitoring of the postoperative patient

    (a) The postoperative neonatal or pediatric patient requires close and continuous monitoring of vital signs and markers of end-organ perfusion. Depending on the preoperative physiologic state, operative intervention, and expected postoperative course, monitoring may be noninvasive or invasive. In addition, the intensivist must be capable of increasing the degree of monitoring based on postoperative dynamic changes. Certain physiologic parameters are monitored postoperatively on a continuous basis (i.e., pulse), whereas others are monitored on an interval basis (i.e., capillary refill). The frequency of interval measurements is dependent on the stability of the patient. For example, a very unstable patient may require interval monitoring to be converted to continuous when applicable (i.e., blood pressure).
    (b) Standard postoperative monitoring includes:

        (i) Cardiac rate and rhythm—continuous
        (ii) Respiratory rate—continuous
        (iii) Pulse oximetry—continuous
        (iv) Capnography—continuous
        (v) Blood pressure via noninvasive oscillometric devices—interval
        (vi) Urine output—continuous via Foley catheter or interval
        (vii) Surgical drain output—continuous or interval
        (viii) Temperature—interval

        (ix) Pain scale—interval
        (x) Blood glucose determinations—interval
        (xi) Weight—daily

    (c) At times, more invasive monitoring may be required and includes:

        (i) Central venous waveform and pressure analysis
        (ii) Intra-arterial waveform and pressure analysis
        (iii) Intracranial waveform and pressure analysis

5. ICU discharge criteria and transition of care to the ward

    (a) Stable gas exchange with decreasing oxygen need and minimal work of breathing
    (b) Stable end-organ perfusion as evidenced by normal hemodynamics and mental status, adequate urine output, resolved acidosis, and stable intravascular volume status
    (c) Stable electrolytes and acid-base status
    (d) Tolerance of enteral nutrition or plan in place to transition from TPN to enteral nutrition
    (e) No ongoing signs of systemic inflammatory response syndrome (SIRS) or sepsis due to presumed infection
    (f) Stable hemoglobin and coagulation profile
    (g) Surgical drain removed or drains with consistently decreasing output
    (h) Adequate pain control on enteral medication or weaning parenteral medications

6. Home discharge criteria

    (a) Consistent clinical features of recovery documented (no ongoing SIRS, tolerance of oral intake, pain controlled on enteral medication, wound healing)
    (b) Outpatient care plan in place and education/training of the family complete. Family confident in assuming medical care to allow completion of convalescence
    (c) Medication education and administration training
    (d) Discharge equipment ordering and training
    (e) Arrangement of home nursing services if needed
    (f) Follow-up appointment arrangements (subspecialists and primary care)

## Study Questions

1. Unlike the adult larynx which is symmetrically cone shaped, a neonate's larynx is…

    a. Tubular with concentric mucosa hypertrophy
    b. Cylindrical with an elliptical cross section and a slightly increased anteroposterior dimension

   c. Shorter, wider, and straighter
   d. More narrow with a significant increase in the lateral dimensions
   e. Nearly identical to adult airways

   Answer to Question 1: (c) Recent MRI and bronchoscopy data reveals the neonatal larynx to be cylindrical in shape with an elliptical cross section and a slightly increased anteroposterior dimension.

2. All the following represent challenges to maintaining functional residual capacity (FRC) in the small infant except…

   a. Infants and neonates lack large cross-sectional areas of the distal airways, creating significant airway resistance in the peripheral small airways.
   b. Branching morphogenesis and alveolar septation are unaltered in preterm infants with severe bronchopulmonary dysplasia.
   c. The chest wall is far more compliant due to a cartilaginous rib cage and poor muscular development both decreasing outward elastic recoil.
   d. Diminished compliance of the lung due to incomplete conducting airway and alveolar development combined with greater interstitial tissue matrix serve to increase inward elastic recoil.
   e. Reduced density of type I muscle fibers and significant respiratory muscle inefficiency at high ventilatory rates increase the risk of early respiratory fatigue.

   Answer to Question 2: (b) Numerous pathologic studies have revealed that severe bronchopulmonary dysplasia in former extremely preterm infants negatively and permanently impacts branching morphogenesis within the lung as well as alveolar septation.

3. The intern calls you to report that a former 24-week infant who needed an inguinal herniorrhaphy requiring general anesthesia at 34-week corrected gestational age remains on the ventilator postoperative and continues to not assist the ventilator. The intern does not understand why the baby has not weaned and is ready for extubation after this relatively short surgical procedure. You relay this important fact to the intern guiding clinical expectations.

   a. Check a blood gas because central respiratory control of breathing is diminished in preterm infants under anesthesia.
   b. The clearance of anesthetics in preterm infants should be rapid.
   c. Maintenance of normal to low normal carbon dioxide levels while on mechanical ventilation should help prevent periodic breathing or frank apnea in preterm infants.
   d. Consider obtaining a scrotal ultrasound to evaluate for testicular torsion.
   e. Obtain an urgent ultrasound of the brain to evaluate for intraventricular hemorrhage.

Answer to Question 4: (a) Preterm infants already have an immature central respiratory control of breathing which can be further diminished under anesthesia and is associated with a markedly decreased ventilatory response to hypercarbia.

## Further Reading

Bang SR. Neonatal anesthesia: how we manage our most vulnerable patients. Korean J Anesthesiol. 2015;68(5):434–41. https://doi.org/10.4097/kjae.2015.68.5.434.

Hudgins PA, Siegel J, Jacobs I, Abramowsky CR. The normal pediatric larynx on CT and MR. Am J Neuroradiol. 1997;18(2):239–45.

# Nutrition

Alfred P. Kennedy Jr and Bethany J. Beaver

**Key Points**

1. Acutely ill surgical patients are at a heightened risk of developing malnutrition.
2. Pediatric malnutrition not only affects growth and development but also negatively impacts surgical outcomes.
3. Enteral nutrition is the preferred route of feeding over parenteral nutrition.
4. Appropriate initiation, advancement, and monitoring are essential when providing safe parenteral nutrition support.

## Introduction

Optimal nutrition not only supports appropriate growth and development but also significantly impacts surgical outcomes. For this reason, the nutrition status of pediatric surgical patients should be assessed and monitored for adequacy. Timely interventions including calorie and protein supplementation, vitamin and mineral additives, and enteral and/or parenteral nutrition support should be considered for best care.

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1) Malnutrition among surgical pediatric patients:
    a) Malnutrition or undernutrition results in poor growth and development, immune dysfunction, delayed wound healing, increased risk of infection, and prolonged hospitalizations. Surgical patients are at a heightened risk of malnutrition due to increased metabolic demands, altered substrate utilization, excessive nutrient losses, and/or inadequate nutrient intake.
    b) For this reason, the nutritional status of surgical patients should be assessed and regularly monitored. Diagnostic criteria established by the American Society of Parenteral and Enteral Nutrition (ASPEN) along with the Academy of Nutrition and Dietetics (AND) can be used to identify malnutrition. Indicators for diagnosis include the following: weight-for-length z-score, BMI-for-age z-score, length/height-for-age z-score, mid-upper arm circumference z-score, weight gain velocity, and nutrient intake.
    c) On growth charts, z-scores provide a more precise means of assessing trends. Z-scores are standard deviations above or below the median (50th%ile) for age and sex. Children who are growing normally will typically fall within the z-scores of $-1$ and 1. Values outside this range should be further assessed for nutritional deficiencies.

2) Nutritional assessment, intake:
    a) Nutritional adequacy can be assessed by comparing current intakes (oral, enteral, and/or parenteral) to usual or baseline intakes. Adequacy can also be determined by comparing current intakes to estimated requirements, especially in the presence of increased metabolic demand.

3) Nutritional assessment, physical findings:
    a) Physically evaluating the fat mass and muscle composition of pediatric surgical patients provides additional information to support a malnutrition diagnosis.
    b) The patient's skin, hair, nails, eyes, teeth, and gums are sensitive to micronutrient deficiencies (Table 1).

4) Nutritional assessment, laboratory:
    a) There is no single laboratory parameter used to assess the nutritional status of pediatric patients.
    b) A comprehensive metabolic panel aids in assessing fluid, electrolyte, and acid/base balance. Serum phosphorus and magnesium should also be considered, especially in the presence of malnutrition. Pediatric patients diagnosed with malnutrition are at an increased risk of developing metabolic derangements.
    c) Serum proteins such as albumin, prealbumin, and retinol binding protein should not be used as indicators of nutritional status. These proteins lack specificity and are often skewed by acute illness.

**Table 1** Micronutrient exam: clinical interpretation of nutrition-focused physical exam findings

| | |
|---|---|
| Hair | |
| Alopecia (thin, sparse, patchy) | Iron, zinc, biotin, protein deficiency |
| Color changes, depigmentation, lackluster | Protein-calorie malnutrition, manganese, selenium, copper deficiency |
| Easily plucked, dull, dry | Protein deficiency, malnutrition, essential fatty acid deficiency |
| Corkscrew hair, unemerged coiled hairs | Vitamin C deficiency |
| Eyes | |
| Angular blepharitis (inflammation of eyelids) | Riboflavin, biotin, vitamin B6, zinc deficiency |
| Pale conjunctiva | Vitamin B6, vitamin B12, folate, iron, copper deficiency, anemias |
| Night blindness, dry membranes | Vitamin A deficiency |
| Bitot's spots (white/gray spots on conjunctiva) | Vitamin A deficiency |
| Face | |
| Pallor | Iron, folate, vitamin B12, vitamin C deficiency |
| Hyperpigmentation (usually around the face, neck, hands) | Niacin deficiency |
| Mouth | |
| Soreness, burning | Riboflavin deficiency |
| Angular stomatitis or cheilitis | Riboflavin, niacin, iron, vitamin B6, vitamin B12 deficiency, vitamin A toxicity |
| Lips | |
| Soreness, burning lips, pale | Riboflavin deficiency |
| Tongue | |
| Sore, swollen, scarlet, raw-beefy red tongue | Folate, niacin deficiency |
| Soreness, burning tongue, purplish/magenta | Riboflavin deficiency |
| Smooth, beefy red tongue | Vitamin B12, niacin deficiency |
| Glossitis (sore, swollen, red, and smooth tongue) | Riboflavin, niacin, vitamin B6, vitamin B12, folate, severe iron deficiency |
| Pale tongue | Vitamin B12, folate, iron deficiency |
| Gums | |
| Gingivitis, swollen, spongy, bleeds easily, redness, retracted gums | Vitamin C, niacin, folate, zinc deficiency, severe vitamin D deficiency, excess vitamin A |
| Neck | |
| Thyroid enlargement, goiter | Iodine deficiency |
| Nails | |
| Beau's lines (transverse ridges, horizontal grooves on the nail) | Severe zinc deficiency, protein deficiency, hypocalcemia |
| Muehrcke's lines (transverse white lines) | Malnutrition |
| Koilonychia (spoon-shaped, concave) | Iron, protein deficiency, anemia |

(continued)

**Table 1** (continued)

| | |
|---|---|
| Brittle, soft, dry, weak, or thin | Magnesium deficiency, severe malnutrition; vitamin A and selenium toxicity |
| Skin | |
| Slow wound healing, decubitus ulcers | Zinc, vitamin C, protein deficiency; malnutrition; inadequate hydration |
| Follicular hyperkeratosis | Vitamin A or vitamin C deficiency |
| Seborrheic dermatitis | Biotin, vitamin B6, zinc, riboflavin, essential fatty acid deficiency; vitamin A toxicity |
| Petechiae | Vitamin C or vitamin K deficiency |
| Purpura | Vitamin C, vitamin K deficiency; excess vitamin E |
| Xerosis | Vitamin A, essential fatty acid deficiency |
| Yellow to orange pigmentation | Excessive beta-carotene |
| Skeletal | |
| Demineralization of bone | Calcium, phosphorus, vitamin D deficiency; excess vitamin A |
| Epiphyseal enlargement of wrists, legs, and knees; bowed legs; rickets or osteomalacia; frontal bossing | Vitamin D deficiency |

Source: Adapted from ref. [1]

   d) Specific vitamin and mineral laboratory parameters may be helpful in determining repletion requirements, especially if signs or symptoms of deficiency exist.

5) Nutritional assessment, anthropometrics:

   a) Age- and sex-specific growth charts should be used to appropriately assess anthropometric data. The Fenton 2013 and the Olsen 2010 growth charts can be used when assessing the growth of preterm infants. Infants that are born term or preterm infants that are corrected to term should be assessed by using the World Health Organization (WHO) growth charts. From age 2 to 20 years, growth parameters should be plotted on the Centers for Disease Control (CDC) growth charts.
   b) Weights should be measured regularly and assessed for trends over time. For infants and children, the rate of weight gain should correspond with the trajectory of his/her growth curve. Deviation from a weight-for-age growth percentile or z-score is concerning for malnutrition.
   c) For infants and children up to 24 months of age, length measurements should be obtained using recumbent length boards. Standing height measurements are appropriate at 2 years. If a child over 2 years or an adult is unable to stand, alternative height measurements can be obtained. These may include knee height caliper, arm span, and tibial length measurements. Deceleration of linear growth may reflect long-term nutritional deficiencies and chronic malnutrition. A stature for age z-score $<-3$ should be assessed for severe malnutrition.

  d) Assessing an infant's weight-for-length can be achieved by plotting the parameters on the WHO growth chart up to 24 months. At 24 months, the child's BMI can be assessed on the CDC growth chart. Infants and children with a weight-for-length or BMI-for-age plotting below the 16th percentile (z score $<-1$) should be assessed for malnutrition.
  e) As with weight-for-length and BMI-for-age, a patient's mid-upper arm circumference can be used to assess for malnutrition from age 6 months to 59 months. This indicator is less effected by fluid changes such as edema and anasarca. Measurements should be plotted and trended on WHO growth charts. Z-score measurements $<-1$ should be assessed for malnutrition.

6) Determining nutrient needs:

  a) Term infants require approximately 100–110 kcal/kg per day and 1.5 g/kg per day of protein. Illness and surgery may alter nutrient requirements, and protein needs may increase to 3 g/kg per day. Premature infants typically require 120–130 kcal/kg per day and 3.5–4.5 g/kg per day of protein.
  b) For older children, energy and protein requirements can be estimated using the 1989 Recommended Dietary Allowance as listed below. This method of estimating nutrient needs is based on the requirements of a healthy child, thus may not reflect the needs of an acutely ill patient. Additional methods of estimating energy needs include using predictive equations with activity and stress factors; however, the most accurate method of predicting energy expenditure is by using indirect calorimetry or a metabolic cart study. Protein requirements among hospitalized patients are typically above the RDA (Table 2).
  c) In some clinical scenarios, children may need more or less than baseline nutritional needs (Table 3).

7) Enteral nutrition:

  a) Oral or enteral nutrition is the preferred method of feeding. Breast milk should be considered initially if available. Breast milk contains immunoglobulins, hormones, and growth factors that cannot be replicated by commercial formulas. Breast milk is estimated to average ~20 kcal/oz (0.67 kcal/mL). Intake of 150–180 mL/kg/day should adequately meet term infant needs. For

**Table 2** Recommended dietary allowances

| Category | Age | Energy (kcal/kg/day) | Protein (g/kg/day) |
|---|---|---|---|
| Children | 1–3 years | 102 | 1.2 |
| | 4–6 years | 90 | 1.1 |
| | 7–10 years | 70 | 1 |
| Males | 11–14 years | 55 | 1 |
| | 15–18 years | 45 | 0.9 |
| Females | 11–14 | 47 | 1 |
| | 15–18 | 40 | 0.8 |

**Table 3** Change in metabolic rate in certain disease states

| Disease state | Approximate % change in metabolic rate (REE) |
|---|---|
| Mild starvation | −5 |
| Peritonitis | +5 |
| Long bone fracture | +10 |
| Severe infection | +40 |
| Multiple trauma | +50 |
| Multiple trauma, ventilated | +60 |
| 50% TBSA burn | +100 |
| 70% TBSA burn | +110 |

REE, resting energy expenditure, TBSA, total body surface area. (Source: Adapted from Essentials of Pediatric Surgery. Rowe MI, O'Neill JA, Grosfeld JL, Fonkalsrud EW, Coran AG. Mosby-Year Book, Inc. 1995. Chapter 7, Nutritional Support of the Pediatric Surgical Patient.)

premature infants, breast milk can be fortified with human milk fortifiers to provide additional calories, protein, and micronutrients. Infants with inadequate intake or infants requiring increased calorie provisions may benefit from fortified breast milk using term infant formula powder. Precise recipes are used to manipulate the nutritional composition of feeds.

b) In the absence of breast milk, standard infant formula should be provided. Standard formulas are cow's milk-based with varying ratios of casein to whey. Premature varieties are also available to provide higher levels of key nutrients. Low or reduced lactose formulas may be appropriate for intolerance and acute diarrhea. Extensively hydrolyzed formulas contain free amino acids and small protein peptides. They are lactose-free and contain varying fat blends. They are considered hypoallergenic and can be used for a cow's milk protein allergy. They may also be considered for infants with short or damaged gut, such as necrotizing enterocolitis. Elemental formulas contain only free amino acids. They are lactose-free and contain a blend of long-chain fatty acids as well as medium-chain fatty acids. Failed use of extensively hydrolyzed formulas or severe malabsorptive conditions would warrant the use of an elemental formula.

c) Children and adults unable to maintain or tolerate adequate nutrition by mouth should be initiated on enteral tube feeds. Commercially prepared, nutritionally complete formulas can be used to meet the nutritional requirements of children and adults. Pediatric formulas are generally used from 1 year to 10 years of age; however, adult formulas can be used before age 10 years if nutritionally indicated. Calorie density ranges from 1 kcal/mL to 2 kcal/mL. Polymeric formulas contain intact proteins and are typically cow's milk-based. Most pediatric and adult formulas are lactose-free. Semi-elemental formulas contain small protein peptides but are not considered hypoallergenic. These formulas may be beneficial for patients with gastrointestinal injury, feeding intolerances, and/or malabsorption. Elemental formulas contain free amino acids and are designed for patients with severe feeding intolerance, malabsorption, or food allergies.

d) For infants, children, and adults, feeding regimens should be provided in a matter that is best tolerated. Bolus feeding is most physiologic; however, continuous feeding may be indicated for feeding intolerance or poor absorption. Feedings given via jejunal tube should always be continuously administered. A combination of daytime bolus feeds and nighttime continuous feeds may be indicated in some situations.

8) Parenteral nutrition:

   a) In the absence of a functional gut, parenteral nutrition should be considered. For older children and adults, the American Society for Parenteral and Enteral Nutrition (ASPEN) recommends initiating parenteral nutrition after 7 days. Parenteral nutrition should be considered sooner for malnourished patients or patients at risk of malnutrition. For infants, parenteral nutrition should be initiated in 1–3 days if enteral/oral feeds cannot be resumed. Children should receive parenteral nutrition within 4–5 days. Sooner initiation of parenteral nutrition is recommended in malnourished patients.
   b) Total parenteral nutrition (TPN) orders are often written with the help of a neonatologist, a pharmacist, and/or a dietitian, but the surgeon must have a familiarity with the process. TPN usually starts with a maintenance volume to which nutrients and electrolytes are added to meet the child's nutritional goals.
   c) Dextrose is the carbohydrate substrate in parenteral nutrition. Dextrose provides 3.4 kcal/g. The initial concentration of dextrose is typically 10%. This is increased daily to a maximum of 25% for central lines. For peripheral parenteral nutrition, a maximum of 12.5% for infants <12 months and 10% for children and adults can be administered safely. The minimum glucose infusion rate to prevent hypoglycemia in newborns is typically 4 mg/kg/min. The maximum is 18 mg/kg/min. Children can tolerate glucose infusions up to 10 mg/kg/min, and adolescents can tolerate up to 6 mg/kg/min. All patients initiated on TPN should have a blood glucose measured daily until the goal regimen is achieved.
   d) Protein requirements vary with age and condition. Protein is given as an amino acid solution. Unlike dextrose, it can often be started at or near goal requirements.
   e) Intravenous fat or lipid emulsions provide calories and a source of essential fatty acids. In pediatric patients, fatty acids are especially important for brain development. Lipids are typically started at a rate of 0.5–1 g/kg/day. Lipids can be advanced daily by 0.5–1 g/kg to a goal of 1–3 g/kg/day. Infants and children need a minimum of 0.5–1 g/kg/day of lipid to prevent essential fatty acid deficiency. Lipids are given as a 10% solution providing 1.1 kcal/mL or a 20% solution providing 2 kcal/mL. Triglyceride levels should be monitored daily until goal lipids are provided.

   f) Additional TPN additives include electrolytes, vitamins, trace elements, and certain medications. Electrolytes should be added to the parenteral admixture and monitored until the goal regimen is achieved. Vitamins and trace elements should also be given based on age and needs. Iron is not included in standard trace elements and is not typically given parenterally. Iron dextran is the only formulation compatible with non-lipid-containing parenteral nutrition. Thus, patients on long-term TPN should be supplemented with enteral iron and/or monitored for iron deficiency.

9) Parenteral nutrition complications:

   a) Pediatric surgical patients on prolonged TPN are at heightened risk of developing multiple parenteral nutrition-related complications. Newborns on TPN often develop cholestasis which can be a major source of morbidity. It is manifest by jaundice and later cirrhosis. Direct hyperbilirubinemia and later transaminasemia are the laboratory indicators of cholestasis. Prolonged parenteral nutrition, non-enteral feeding, underlying illness, and recurrent infections increase the likelihood of developing cholestasis and subsequently parenteral nutrition-associated liver disease (PNALD). When feasible, protective measures should be implemented for infants and children requiring prolonged parenteral nutrition. Enteral feeding is the single most important intervention for protecting and correcting PNALD. Parenteral nutrition cycling which limits the duration of TPN throughout the day may also minimize the risk of developing cholestasis; however, cycling may be limited by the patient's age and size. Sudden or prolonged cessation of TPN may result in hypoglycemia. Tapering start-up and stop times may help prevent this from occurring. Additional strategies include reducing lipid provisions and/or choosing an alternate lipid source with increased omega-3 fatty acids. Phenobarbital and ursodiol may help with bile elimination.
   b) Additional complications associated with parenteral nutrition include central line-associated bloodstream infections, electrolyte derangements, vitamin/mineral deficiencies, aluminum toxicity, and metabolic bone disease. Regular monitoring with a skilled team is essential for preventing complications.

## Additional Notes

Pediatric surgical patients are at a heightened risk of developing nutritional deficiencies and malnutrition. Poor nutritional standing has been shown to negatively impact growth, development, and surgical outcomes. Thus, assessing the nutritional status and implementing appropriate interventions should be a standard part of caring for a pediatric surgical patient.

## Study Questions

1. A malnourished patient is at increased risk of which of the following?

   a) Poor wound healing
   b) Increased hospital length of stay
   c) Increased hospital cost
   d) All of the above

   Answer: (d) All of the above. Malnutrition negatively impacts clinical outcomes and increases hospital costs. Malnutrition in pediatric patients is detrimental to growth and development.

2. Which of the following feeding regimens is most appropriate for a 3-month-old infant with a past medical history of poor oral skills now requiring a gastrostomy placement?

   a) Continuous feeds of expressed breast milk at 30 mL/h × 24 h
   b) Bolus feeds of standard 20 kcal/oz infant formula at 120 mL Q4H
   c) Bolus feeds of a 20 kcal/oz extensively hydrolyzed formula at 120 mL Q4H
   d) Continuous feeds of 30 kcal/oz (1 kcal/mL) pediatric formula at 30 mL/h × 24 h

   Answer: (b) Bolus feeds of standard 20 kcal/oz infant formula at 120 mL Q4H. Breast milk is not sterile and should not be run continuously for risk of bacterial contamination. Continuous feeds are also least physiologic and should be avoided unless necessary. If there is no history of allergy or intolerance, a standard infant formula would be preferred over an extensively hydrolyzed or elemental formula. Pediatric formulas should not be used before 12 months of age.

3. A 5-year-old male is readmitted to the hospital following appendectomy POD#6 with multiple days of N/V and concern for septic abdomen. The most appropriate nutritional intervention for this patient would be which of the following?

   a) Clear liquids for 3–4 days.
   b) NPO, initiate parenteral nutrition.
   c) Resume a pediatric regular diet.
   d) Start trophic enteral feeds with an elemental formula.

   Answer: (b) NPO, initiate parenteral nutrition. Bowel rest and parenteral nutrition would be the most appropriate intervention based on the duration of his N/V and predicted poor nutritional intake. Enteral nutrition would be contraindicated.

4. To minimize the risk of cholestasis, which intervention would prove most beneficial?

   a) Extend the duration of TPN over 24 h per day to minimize the glucose infusion rate.
   b) Reduce protein provisions and subsequently increase lipid provisions to meet goal calorie requirements.
   c) Add famotidine to the TPN.
   d) Initiate enteral feeding.

   Answer: (d) Initiate enteral feeding. Enteral feeding alone is the single most protective measure against the development of cholestasis. Cycling TPN and reducing lipids may provide some benefit as well.

## Reference

1. Mordarski B, Wolff J. Pediatric nutrition focused physical exam pocket guide. Ac Nut Diet. 2018

## *Further Reading*

Baylor College of Medicine Section of Neonatology, Department of Pediatrics. Guidelines for Acute Care of the Neonate. 26th Edition. 2018–2019

Becker PJ, Nieman Carney L, Corkins MR, et al. Consensus statement of the academy of nutrition and dietetics/American Society for Parenteral and Enteral Nutrition: indicators recommended for the identification and documentation of pediatric malnutrition (undernutrition). J Acad Nutr Diet. 2014;114(12):1988–2000.

Corkins MR, Balint J. The A.S.P.E.N. pediatric nutrition support core curriculum. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2015.

Grover Z, Ee LC. Protein energy malnutrition. Pediatr Clin N Am. 2009;56(5):1055–68.

Mehta NM, Corkins MR, Lyman B, et al. Defining pediatric malnutrition. J Parenter Enter Nutr. 2013;37(4):460–81.

Recommended Dietary Allowances, 10th ed, National Academy of Sciences, National Academy Press 1989; 33–36.

Worthington P, Balint J, Bechtold M, et al. When is parenteral nutrition appropriate? J Parenter Enter Nutr. 2017;41(3):324–77.

# Disorders of Clotting

**Michal A. Miller**

**Key Points**

1) Disorders of clotting may be hereditary illnesses that children carry with them their entire lives, such as hemophilia, or they may be temporary conditions due to illness, organ failure, medication, or trauma.
2) Some disorders of clotting will not be known until an operation has already started. This is often the case with previously undiagnosed von Willebrand's disease, which is the most common hereditary bleeding disorder.
3) Safe performance of an operation may require correction of deficient clotting factors and platelet count. The expert consultation of a pediatric hematologist will improve safety in complex cases.

Clotting dysfunction is of particular interest to surgical specialists as it can drastically alter the intraoperative and postoperative course and lead to hemorrhagic complications. Some bleeding disorders will be known to the child's family because they were previously diagnosed. Preoperative testing, although rarely indicated in a healthy child, can detect temporary and situational disorders of clotting and allow for correction prior to an operation. Some disorders of clotting will only be discovered once the operation has started, and the surgeon must be able to respond to unexpected bleeding.

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1. Evaluation of clotting function
    a. Family history, the patient's history of bleeding, including site of bleeding and severity of the abnormal bleeding, age, and prior surgical complications.
    b. Of note, the possibility of child abuse should be considered in any child with abnormal bleeding and/or bruising. Recurrent bleeding at specific site generally indicates a local anatomical cause or recurrent local trauma. Bruising over bony prominences such as the pre-tibial areas is common in active children. Large bruises over soft tissue area such as the abdomen raise the suspicion of abuse or a bleeding disorder [1].
    c. Laboratory evaluation
        i. Complete blood count (CBC) is the initial laboratory evaluation. A platelet count of >100,000 is usually clinically asymptomatic and frequently does not result in significant intraoperative bleeding. The presence of iron deficiency anemia (microcytic hypochromic anemia) can be a sign of unappreciated chronic bleeding. Pancytopenia signals a need for further evaluation.
        ii. Coagulation studies, to include prothrombin time (PT), international normalized ratio (INR), and activated partial thromboplastin time (aPTT).
        iii. Platelet function assay (PFA) has replaced bleeding time as a screen for platelet function.
        iv. Fibrin split products (FSP)/fibrinogen
        v. Clotting factor deficiencies other than Factor VIII and Factor IX are rare. A hematologist should be consulted for evaluation of specific factor deficiencies.
    d. Clinical presentation
        i. Clotting factor deficiencies: Most common presenting signs are bleeding into joints, soft tissues, or muscles. The visible bruising is frequently large, deep palpable, and located on the trunk as well as the extremities. The degree of bruising is out of proportion to the degree of trauma. Frequently the bruising occurs spontaneously without a history of trauma. The increased surgical bleeding is usually more impressive postoperatively than intraoperatively. Small superficial cuts do not tend to bleed excessively.
        ii. Platelet dysfunction and thrombocytopenia: Most common presenting signs are petechiae, frequent superficial bruising, and mucous membrane bleeding such as epistaxis, menorrhagia, and gingival bleeding. The increased surgical bleeding is usually more impressive intraoperatively than postoperatively. Small superficial cuts tend to bleed excessively. Platelet dysfunction disorders present similar to thrombocytopenia.
        iii. Thrombocytopenia. The degree of bleeding is proportional to the counts. A platelet count of >100,000 is usually not associated with excessive

bleeding. 50,000–100,000 results in a moderate increase and 20,000–50,000 in severely increased bleeding complications. Spontaneous bleeding usually does not occur until the count is <20,000.

2. Hereditary bleeding disorders
    a. Hemophilia A: Factor VIII deficiency and an elevated aPTT. Incidence of hemophilia A in healthy newborns is approximately 1:5000. It has X-linked inheritance. Approximately one-third of severe cases are from spontaneous mutations and have no family history.
    b. Hemophilia B: Factor IX (Christmas factor) deficiency. Incidence of hemophilia B is less common than factor VIII deficiency and comprises approximately 15% of cases of hemophilia. It has X-linked inheritance.
    c. Hemophilia C: Factor XI deficiency. It is rare and has autosomal recessive inheritance.
    d. Severity of disease
        i. Severe hemophilia is characterized by a factor level of less than 1%. Bleeding occurs spontaneously and results in long disability without prophylactic administration of factor VIII.
        ii. Moderate hemophilia is characterized by factor level of 1–5%. Prolonged bleeding occurs after mild to moderate injuries but does not usually occur spontaneously.
        iii. Mild hemophilia is characterized by factor level of 5–10%. Mild hemophilia can go undiagnosed until the time of operation.
    e. Von Willebrand's disease
        i. Von Willebrand's disease (vWD) is the most common hereditary bleeding disorder. Coagulation studies are normal in this condition. The incidence is reported to be as high as 2% of the general population. vWD is the result of a deficiency in and/or dysfunction of von Willebrand factor (vWF). vWF is involved in adherence and aggregation of platelets in response to damaged endothelium. It also binds to plasma factor VIII and increases the half-life of plasma factor VIII. It is synthesized in megakaryocytes and endothelial cells. vWF is stored within the Weibel-Palade bodies in endothelial cells and within the alpha granule in platelets. It is frequently not diagnosed until operation when bleeding occurs in a child who has a history of easy unusual bruising, frequent epistaxis, or menorrhagia.
        ii. Classification
            1. Type 1 is a reduction in the amount of vWF. Type 1 is the most common type of vWD and accounts for approximately 75% of cases. Inheritance is autosomal dominant with variable penetrance. Affected family members of the patient can have different clinical expression. The clinical severity is primarily dependent on the degree of deficiency, but other endogenous as well as exogenous factors can influence the

    severity of bleeding. The deficiency results in increased bruising, menorrhagia, mucosal bleeding, and bleeding during operation.
    2. Type 2 is a group of subtypes that consist of dysfunctional vWF activity. This group is uncommon, and inheritance can be dominant or recessive.
    3. Type 3 is severe reduction or absence of vWF. It is rare. Inheritance is recessive. If VWF is absent or markedly reduced, clinical symptoms are similar to hemophilia A with bleeding into muscles and joints.

3. Acquired pathological bleeding disorders
    a. Liver disease/injury results in decreased hepatic synthesis of clotting factors and increases fibrinolytic activity.
    b. Kidney disease can lead to loss of coagulation factors, and uremia can decrease platelet function.
    c. Gastrointestinal malabsorption results in vitamin K deficiency and deficiency of vitamin K-dependent coagulation factors (II, VII, IX, and X). The PT and INR will be elevated in this deficiency.
    d. Disseminated intravascular coagulation (DIC) results in consumption and activation of multiple coagulation and anticoagulation factors. Laboratory studies will show a low platelet count, elevation of D-dimer concentration, and decreased fibrinogen concentration. CBC may show schistocytes, which result when fibrin strands in clot damage erythrocytes.
    e. Hypersplenism causes moderate thrombocytopenia from sequestration. Conversely, splenectomy will lead to thrombocytosis with platelet counts frequently in the 400,000–600,000 range.
    f. Cyanotic congenital heart disease results in decreased platelet aggregation and shortens platelet survival.

4. Therapy
    a. Treatment of hemophilia A and B: Factor VIII and IX recombinant factors are widely available for replacement. It is preferable to use the same brand of factor the patient has been using. This limits complications.
    b. Treatment vWD
        i. Desmopressin (DDAVP) stimulates the release of VWF from storage sites into plasma. Repeated doses deplete the storage pool, and thus DDAVP is generally not administered for more than two doses. If a sustained level of vWF is needed for a longer period of time, or if the patient has type 2 or 3 vWD, DDVP will be ineffective and factor replacement is needed. Of note, repeated doses of DDAVP can lead to hyponatremia.
        ii. vWF replacement: Recombinant vWF is not widely available. Currently, anti-hemophilic factor VIII concentrates that also contain vWF are the primary source for vWF replacement.

c. Transfusion
    i. Platelet transfusion
        1. Conservative platelet transfusion indications based on platelet count/scenario:
            a. < 20,000: immediate prophylactic.
            b. <30,000 with minor bleeding (epistaxis, mucosal bleeding menorrhea, or persistent "oozing" from incision).
            c. <50,000 in patients with major trauma, CNS injury, or newly diagnosed cancer.
            d. <50,000 prior to minor procedures such as a bone marrow aspiration or lumbar puncture.
            e. <75,000–100,000 prior to a minor operation such as central venous line placement or gastrostomy tube placement.
            f. <100k prior to major operation and neurosurgical operations.
            g. If the operation is prolonged, additional transfusion may be needed.
        2. Single donor apheresis units are now standard of care. Random donor units can be used if they are all that is available. A transfusion of 10–15 ml/kg (or 1 unit for an adult-sized child) is expected to increase the platelet count by 50,000–100,000. Crossmatch additional units to transfuse in case bleeding arises intraoperatively [2].
    ii. Packed red blood cell (pRBC) transfusion: 10 ml/kg (or 1 unit for an adult-sized child) is expected to increase the hemoglobin by 1.0–1.5g. Crossmatch additional units to transfuse if bleeding arises intraoperatively.
    iii. The standard of care for type 1 vWD and hemophilia A or B is factor replacement via commercially available factor concentrates. In an emergency when factor replacement is not available, fresh frozen plasma (FFP) can be used. In general, the degree and duration of factor replacement is dependent on the surgical procedure.
    iv. Rare clotting factor deficiencies require FFP.
    v. Platelet dysfunction disorders frequently are not thrombocytopenic but require platelet transfusion to provide functional platelets.
    vi. Special situations: Irradiated leukoreduced blood products should be used for patients with an underlying congenital immunodeficiency, infants < 3 months old, and iatrogenic immunodeficiency such as chemotherapy and transplant patients.
    vii. Patients who have received multiple transfusions can become refractory. Ill patients can consume platelets. Hypersplenic patients will sequester platelets. This occurs in patients with immune thrombocytopenia such as idiopathic thrombocytopenic purpura (ITP).

- viii. FFP contains both pro- and anticoagulation factors. In general, it is the agent of choice to rapidly replace patients with multiple factor deficiencies as occurs in massive hemorrhage or when there is a need for urgent reversal of therapeutic warfarin anticoagulation. FFP is also a source of factors for patient with rare congenital factor deficiencies for which factor concentrates are not yet available.
- ix. Cryoprecipitate most importantly contains fibrinogen. Historically, it was a source of vWF and factor VIII. Today, patients with hemophilia and vWD should receive commercially available factor concentrates. Activated factor VII is indicated to treat hemorrhage in hemophilia patients who have antibodies to factor VIII or factor IX.
- x. Commercially available hemostatic agents
    1. Thrombin
    2. Geofoam (Pfizer, Franklin, OH)
    3. Fibrin glue and sealants
    4. Surgicel (Johnson & Johnson, New Brunswick, NJ)
    5. QuikClot (Teleflex, Wayne, PA)

5. Complications of transfusion
    - a. Hepatitis B transmission: incidence of 1 in 200,000 to 500,000
    - b. Hepatitis C transmission: incidence of 1 in 500,000
    - c. Human immunodeficiency virus (HIV) 1 and 2: incidence of <1 in 1 million
    - d. Bacterial infection can be as high as 1 in 1000. Usually occurs with platelet transfusion
    - e. Transfusion-related acute lung injury (TRALI)
    - f. Transfusion-associated circulatory overload (TACO)
    - g. Acute/delayed hemolytic transfusion reactions from antibodies against RBC antigens
    - h. Iron overload
    - i. Fluid overload
    - j. Febrile non-hemolytic transfusion reaction
    - k. Acute onset hemolytic reaction
    - l. Delayed transfusion reaction: 2–14 days after a transfusion
    - m. Alloimmunization in females who are Rh negative
    - n. Sepsis from bacterial contamination
    - o. Allergic reaction
6. Clotting cascade (Fig. 1). The extrinsic pathway affects PT, the intrinsic pathway affects aPTT, and the common pathway affects thrombin time.

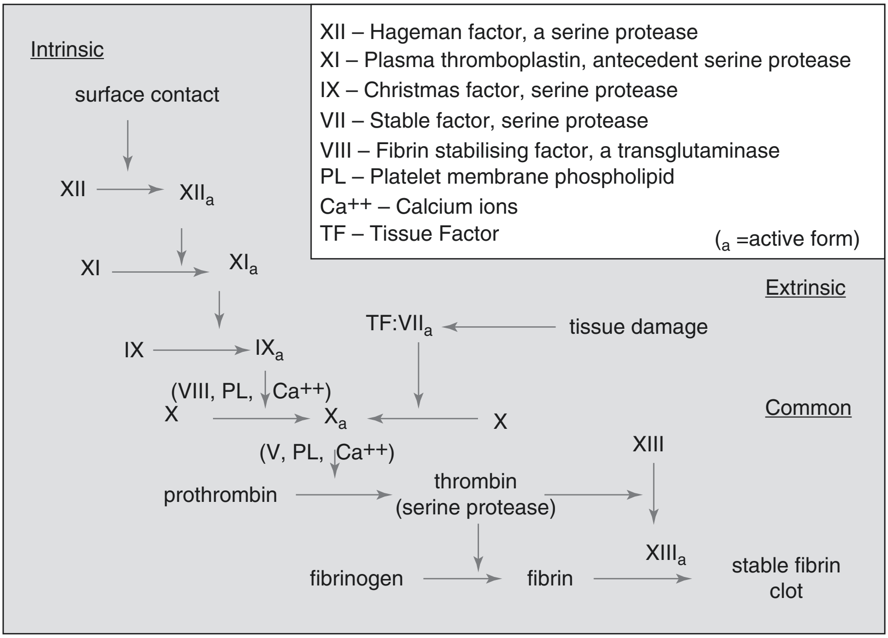
XII – Hageman factor, a serine protease
XI – Plasma thromboplastin, antecedent serine protease
IX – Christmas factor, serine protease
VII – Stable factor, serine protease
VIII – Fibrin stabilising factor, a transglutaminase
PL – Platelet membrane phospholipid
Ca++ – Calcium ions
TF – Tissue Factor
(a =active form)
Intrinsic
surface contact
XII
XIIa
XI
XIa
IX
IXa
(VIII, PL, Ca++)
X
Xa
X
Extrinsic
TF:VIIa
tissue damage
Common
(V, PL, Ca++)
prothrombin
thrombin
(serine protease)
XIII
XIIIa
fibrinogen
fibrin
stable fibrin clot

**Fig. 1** The clotting cascade: The three pathways that make up the classical blood coagulation pathway are the intrinsic, extrinsic, and common pathways (Adapted from GrahamColm under Creative Commons Attribution-Share Alike 3.0 Unported license as published on https://en.wikipedia. https://commons.wikimedia.org/wiki/File:Classical_blood_coagulation_pathway.png, Accessed 28 Dec 2013)

## Additional Notes

Disorders of clotting create a challenge to the safety of a child who needs to undergo an operation. Most problems can be corrected, albeit temporarily, with the administration of blood products, deficient clotting factors, or medications to make clotting function sufficient to safely pass through an operation. It is often helpful to consult the expertise of a pediatric hematologist to assist with specific clotting factor replacement and protection of the child from postoperative bleeding.

A modality that is broadly available to determine specific deficiencies in the formation of clot is thromboelastography (TEG) (Fig. 2). TEG can help guide which specific step in the formation of clot is not occurring properly (Table 1 and Fig. 3).

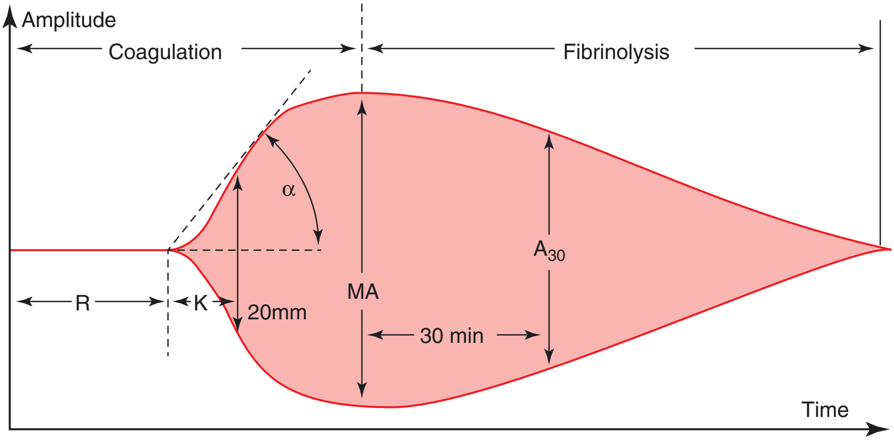

**Fig. 2** Parameters in thromboelastography (TEG). R: reaction time to initial fibrin formation. K: kinetic time for fibrin cross linkage to reach 20 mm clot strength. α: angle from baseline to slope of viscosity (clotting). MA: maximum amplitude of clotting. A30: amplitude of tracing at 30 min after MA. (Source: https://commons.wikimedia.org/wiki/File:Thromboelastography_parameters.png , Accessed 4 Nov 2021, Used under Creative Commons CC0 1.0 Universal Public Domain Dedication)

**Table 1** Indicated treatment for abnormalities in TEG (thromboelastography) values

| TEG value | Normal range | Function measured | Indicated treatment if abnormal |
|---|---|---|---|
| TEG-ACT | 80–140 s | Intrinsic/extrinsic path | >140 s: FFP |
| R time | 5–10 min | Intrinsic path | >10 min: FFP |
| K time | 1–3 min | Fibrinogen, platelet # | >3 min: cryoprecipitate |
| α angle | 53–72 ° | Fibrinogen, platelet # | <53°: cryoprecipitate, platelets |
| MA | 50–70 mm | Platelet # and function | <50 mm: platelets |
| A30 | 0–3% loss | Fibrinolysis | >3%: tranexamic acid |

TEG-ACT: activated clotting time to initial formation of fibrin. R: reaction time to initial fibrin formation. K: kinetic time for fibrin cross linkage to reach 20 mm clot strength. α: angle from baseline to slope of viscosity (clotting). MA: maximum amplitude of clotting. A30: amplitude of tracing at 30 min after MA. (Adapted from ref. [3])

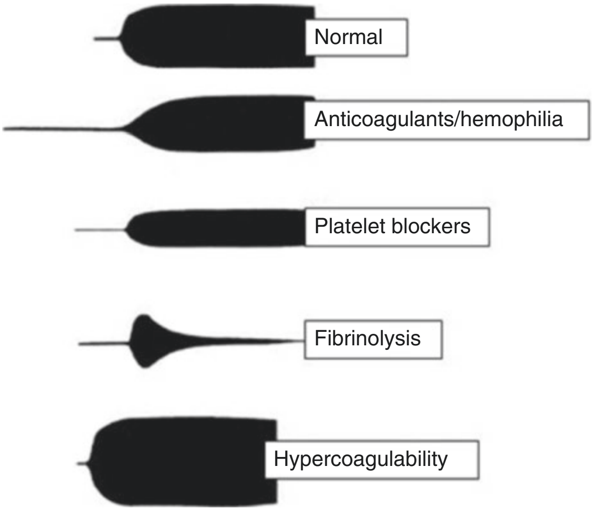

**Fig. 3** Abnormal patterns of thromboelastography (TEG) in states of disordered clotting. The tracing of the TEG viscosity curve will deviate from the normal pattern, seen at the top of the figure, when clot function is abnormal. The values measured during TEG will guide which therapy is indicated. (Source: Teodoro Da Luz L, Nascimento B, Rizoli S. Thromboelastography (TEG®): Practical considerations on its clinical use in trauma resuscitation. Scand J Trauma Resusc Emerg Med. 21(1): 29, 2013. https://pubmed.ncbi.nlm.nih.gov/23587157/, Accessed 4 Nov 2021. Used under CC BY 2.0 License, https://creativecommons.org/licenses/by/2.0/)

## Study Questions

1) Question 1: What is the most common inherited bleeding disorder?

   a. Hemophilia A
   b. Hemophilia B
   c. Von Willebrand's disease (vWd)
   d. Vitamin K deficiency

   Answer: (c.) Von Willebrand's disease (vWd). Von Willebrand's disease (vWD) is the most common hereditary bleeding disorder. The incidence is reported to be as high as 2% of the general population. vWD is the result of a deficiency in and/or dysfunction of von Willebrand factor (vWF). vWF is involved in adherence and aggregation of platelets in response to damaged endothelium. It also binds to plasma factor VIII and increases the half-life of plasma factor VIII. It is synthesized in megakaryocytes and endothelial cells. vWF is stored within the Weibel-Palade bodies in endothelial cells and within the alpha granule in platelets. It is frequently not diagnosed until operation when bleeding occurs in a child who has a history of easy unusual bruising, frequent epistaxis, or menorrhagia.

2) Question 2: Which clotting factor is deficient in hemophilia B?

   a. Factor VII
   b. Factor IX (Christmas factor)
   c. Factor XI
   d. Vitamin K

   Answer: (b.) Factor IX (Christmas factor). Hemophilia A is a bleeding disorder characterized by factor VIII deficiency. Incidence of hemophilia A in healthy newborns is approximately 1:5000. It has X-linked inheritance. Approximately one-third of severe cases are from spontaneous mutations and have no family history. Hemophilia B is a deficiency of factor IX (Christmas factor). Incidence of hemophilia B is less common than factor VIII deficiency and comprises approximately 15% of cases of hemophilia. It has X-linked inheritance. The rarest form of hemophilia is hemophilia C, factor XI deficiency. It is rare and has autosomal recessive inheritance.

3) Question 3: A 6-year-old female has been newly diagnosed with leukemia. She requires placement of a Mediport so that she may begin chemotherapy. Complete blood count (CBC) reveals that she has a white blood count of 3.8 K/uL, hemoglobin of 7.4 g/dL, and a platelet count of 12 K/uL. What is the appropriate next step in her treatment?

   a. Cancel operation, discharge child home, and follow up in clinic in a month to re-test CBC.
   b. Proceed with operation without change in plan.
   c. Transfuse packed red blood cells until hemoglobin concentration reaches 10 g/dL.
   d. Transfuse platelets and proceed with operation when level is adequate (typically 75 K/uL, but this threshold may vary between surgeons).

   Answer: (d.) Transfuse platelets and proceed with operation when level is adequate (typically 75 K/uL, but this threshold may vary between surgeons). This child requires central access of some sort to initiate treatment of her newly diagnosed leukemia. It is not an option to delay this operation. A hemoglobin level of 7 g/dL or greater is acceptable for a minor operation; she does not need transfusion of packed red blood cells. Proceeding with the operation at her level of thrombocytopenia puts her at risk for bleeding complications which may be life-threatening. The safest course of therapy is to transfuse platelets until her platelet level reaches at least 75 K/uL and then perform placement of the Mediport so that chemotherapy may be initiated.

## References

1. Carpenter SL, et al. Evaluating for suspected child abuse: conditions that predispose to bleeding. Pediatrics. 2013;131(4):e1357–73.
2. Callow CR, et al. The frequency of bleeding complications in patients with haematological malignancy following the introduction of a stringent prophylactic platelet transfusion policy. Br J Haematol. 2002;118:677–82.
3. Simon E, Koyfman A, Long B. The Thromboelastogram (TEG®): a five-minute primer for the emergency physician. emDocs, 2016. http://www.emdocs.net/thromboelastogram-teg-five-minute-primer-emergency-physician/. Accessed 4 Nov 2021.

## *Further Reading*

Orkin SH. Nathan and Oski's hematology of infancy and childhood. 7th ed. Philadelphia, PA: Saunders Elsevier; 2009.

Teodoro Da Luz L, Nascimento B, Rizoli S. Thromboelastography (TEG®): Practical considerations on its clinical use in trauma resuscitation. Scand J Trauma Resusc Emerg Med. 2013;21(1):29.

# Part III
# Trauma

# Epidemiology of Trauma

**Bryan S. Walters, Emily E. Steinhilber, and Christopher P. Coppola**

**Key Points**

1. Trauma is the leading cause of death for children. It is the prime reason for years of life lost and medical costs in the care of children.
2. Homicide and suicide are more common causes of death for teenagers.
3. Abuse is often a lethal disease for infants.
4. Head injury mixed with another traumatic injury results in an increased chance of death.
5. Males are more likely than females to have injuries involving higher energy transfer, while less likely to use protective devises. They are also more likely to be injured deliberately.
6. Unintentional falls are the leading cause of nonfatal injuries.

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1. Epidemiology by body region
    a. Head
        i. In ages less than 2 years old, physical abuse is the most common cause of head injury.
        ii. A child's brain doubles in size during the first 6 months of life and reaches 80% of an adult's brain size by 2 years old.
        iii. In children 3 years old and above, the three most common head injury mechanisms are:
            1. Motor vehicles
            2. Bicycle crashes
            3. Falls
        iv. Children are five times more likely to have apnea, hypoventilation, and hypoxia from blunt trauma to the brain than they are to have hypovolemia-associated hypotension. Pulmonary failure is common; Cardiac failure is uncommon and is usually secondary to pulmonary failure.
    b. Cervical spine
        i. Cervical spine injuries comprise 80% of spinal trauma in pediatrics. Children are 2.5 times more likely than adults to suffer a C-spine injury due to large head size and a highly flexible spine.
        ii. Ligamentous injuries are more common than fractures.
        iii. Hyperflexion injuries (anterior wedge fractures and posterior disruption) are more common than hyperextension (compression of posterior and anterior disruption of the longitudinal ligament).
        iv. The upper cervical spine (C1–C3) is the most vulnerable site in children under 8 years old.
        v. Symptoms are delayed in up to 25% of children with cervical spine injuries, with symptoms typically presenting 30 min to 4 days later.
        vi. Pseudo-subluxation of C2 on C3 is a normal finding for up to 38% of children under 7 years old and in 20% of children up to 16 years old.
        vii. Children are more likely than adults to sustain spinal cord injury without radiographic abnormality (SCIWORA). A normal cervical spine series may be present in two-thirds of children with spinal cord injuries.
    c. Thoracic
        i. Occurs infrequently in children: Approximately 5–12% of pediatric trauma patients. However, thoracic injuries are second only to head trauma as the most common cause of death for these patients.

    ii. Around 85% of thoracic trauma cases are due to blunt forces, with motor vehicle accidents as the most common source.
    iii. Penetrating thoracic injuries are less common yet carry a 15% mortality rate.
    iv. Pulmonary contusion is the most common thoracic injury in a child.
    v. More than 80% of children with pulmonary contusions also have extrathoracic injuries; thus, mortality rates for children with pulmonary contusions can be as high as 22%.

d. Abdominal

    i. Eighty percent of abdominal injuries are due to blunt force.
    ii. Isolated abdominal injuries are most common.
    iii. The spleen is the most commonly injured abdominal organ.
    iv. Solid organ injuries of the abdomen typically occur by one of two mechanisms: direct blow or high energy injuries (motor vehicle, all-terrain vehicle, and fall from a great height).
    v. Renal injuries in children are less common (rare under age 5 years old) due to anatomical differences, seen in approximately 10% of all pediatric blunt abdominal trauma.
    vi. Multi-system abdominal injuries carry mortality as high as 12%.

e. Pelvic

    i. Most commonly related to high-energy mechanisms. Pedestrian struck by motor vehicle is the most common mechanism.
    ii. Occur less frequently in children versus adults (1.6% of all pediatric trauma admissions).
    iii. Children have a lower mortality rate from pelvic hemorrhage than adults due to anatomical differences.
    iv. Pelvic ring disruption and avulsion injuries are more commonly reported as they can occur with lower energy impacts.
    v. Injuries commonly associated with pelvic injuries:
        1. Hemorrhage from other injuries
            a. Up to 30% of patients require blood transfusion(s) due to hemorrhage from an associated injury.
        2. Head injury: Up to 40%
        3. Chest/abdomen: 15–33% association
        4. Genitourinary: 11–12%
        5. Neurologic: Up to 6%
        6. Extremity: 18% (femur and tibia most common) (Fig. 1)

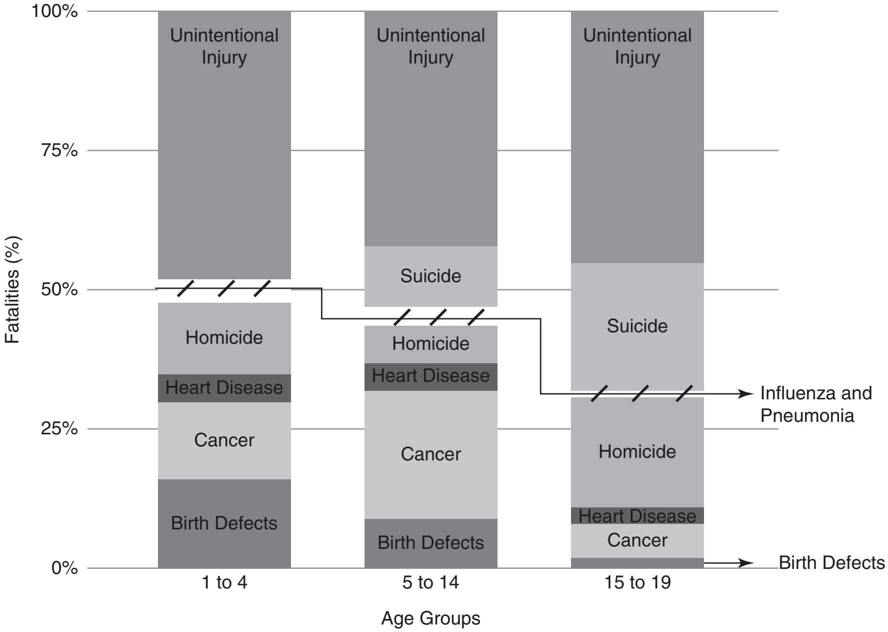

**Fig. 1** Pediatric fatality by type and age. (Source: Emily Steinhilber)

## Additional Notes

While epidemiology can be a dry subject, the data tracked by trauma registries is highly useful. Epidemiologic information pertaining to trauma has helped direct public health initiatives, thus helping countless children. Initiatives that focus on prevention will save far more lives than those that focus on treatment.

Advanced trauma life support (ATLS) guidelines are essential in the care of pediatric trauma patients, to try and make as many as possible of pediatric injuries survivable. Communication between pre-hospital providers and Trauma Center staff is key to maximize the opportunity to reverse correctible threats to life during the initial “Golden Hour” after injury.

Understanding regional differences in trauma epidemiology is useful in the evaluation of the pediatric trauma patient. For example, nine out of ten children under age 15 that are killed by firearms live in the United States. Differences in regional injury patterns are seen when comparing one geographic terrain to another, e.g., one city to another or urban versus rural. This data can be used to guide transfer and treatment decisions at any Trauma Center.

## Study Questions

1) The leading cause of death in children is ______.

   a) Splenic injuries
   b) Acute lymphocytic leukemia
   c) Trauma
   d) Cancer

   Answer: (c) Trauma. For children, trauma is a greater cause of mortality than any other etiology for all ages. It is greater cause of mortality than the next three causes of death combined.

2) Children are 2.5 times more likely than adults to suffer a/an ______ injury due to large head size and a highly flexible spine.

   a) Cervical spine
   b) Liver
   c) Thoracic spine
   d) Ophthalmic

   Answer: (a) Cervical spine. In children, in particular infants and small children, the head represents a greater portion of body surface area and weight as compared to adults. Blunt traumas that involve deceleration cause the weight of the head to pitch forward or back and exert force on the cervical spine.

3) ______ abdominal injuries carry a mortality rate as high as 12%.

   a) Pediatric
   b) Multi-system
   c) Missed
   d) Unfounded

   Answer: (b) Multi-system. When a child suffers multi-system trauma, the risk of death is greater than if any one system is injured alone. Head injury combined with hemorrhagic shock is particularly lethal.

4) Under the age of 2 years old, ______ is the most common cause of head injuries.

   a) Fall
   b) Physical abuse
   c) Motor vehicle collision
   d) All-terrain vehicle accident

   Answer: (c) Physical abuse. Providers who treat children must have a high index of suspicion for non-accidental trauma. As the history of the trauma can be absent or erroneous, non-accidental trauma can be missed if not actively investigated. Children under the age of 2 years old are not typically involved in activities involving high forces of energy, as is more common in adolescents. Severe head injury in a young child requires consideration of child abuse as an etiology, especially if there is no clear explanation of how the injury was caused.

## Further Reading

Fowler KA, Dahlberg LL, Haileyesus T, Gutierrez C, Bacon S. Childhood firearm injuries in the United States. Pediatrics. 2017;140:1–3.

Grinshteyn E, Hemenway D. Violent death rates: the US compared with other high-income OECD countries, 2010. Am J Med. 2016;129(3):266–73.

Grinshteyna E, Hemenway D. Violent death rates in the US compared to those of the other high-income countries, 2015. Prev Med. 2019;123:20–6.

Meagher AD, Zar zaur BL. Epidemiology. In: Feliciano DV, Mattox KL, Moore EE. eds. Trauma, 9e. McGraw-Hill; Accessed April 14, 2021. https://accesssurgery.mhmedical.com/content.aspx?bookid=2952&sectionid=249116193

NTDB Reports and Publications. 2021. https://www.facs.org/quality-programs/trauma/tqp/center-programs/ntdb/docpub. Accessed 14 Apr 2021.

Pediatric TQIP: An overview. 2021. https://www.facs.org/quality-programs/trauma/tqp/center-programs/tqip/pediatric-tqip. Accessed 14 Apr 2021.

WISQARS (web-based injury STATISTICS query and Reporting System) | Injury CENTER | CDC. (2020, July 01). https://www.cdc.gov/injury/wisqars/index.html. Accessed 14 Apr 2021.

# Injury Prevention

**Judith Egly**

**Key Points**

1. Injury is the most common cause of death for children in all age groups. Prevention of injury in children will save lives, prevent disability, decrease time spent in the hospital, and lower the cost of health care costs.
2. Injury prevention begins in the home with measures as simple as child-proofing but and extends to broader efforts including education, advocacy, helmets, car seats, education, safety fairs, regulations, and legislation.
3. Several organizations work to provide support and guidelines to families and providers striving to reduce injury in children. These include Safe Kids Worldwide, the American Academy of Pediatrics, the Consumer Product Safety Council, and the Centers for Disease Control and Prevention to name a few.

For more than 10 years, preventable injuries have been the leading cause of death for children ages 1–19 according to the Centers for Disease Control and Prevention's (CDCP) National Center for Injury Prevention and Control (2008–2018) (Fig. 1).

In 2019, more than 7000 children died in the United States due to unintentional injury, which equates to 20 children each day [1]. The famous quote from C. Everett Koop, MD, former Surgeon General, highlights the significance of this statistic. He said, "If a disease were killing our children in the proportions that 'accidents' are, people would be outraged and demand that this killer be stopped. Most injuries to people-and nearly all injuries to children-can be predicted and can be prevented."

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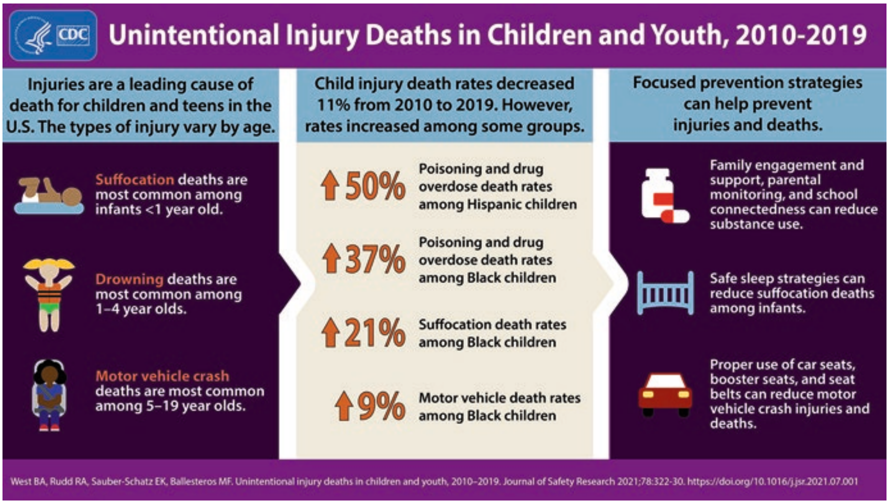

Unintentional Injury Deaths in Children and Youth, 2010-2019

Injuries are a leading cause of death for children and teens in the U.S. The types of injury vary by age.

Suffocation deaths are most common among infants <1 year old.

Drowning deaths are most common among 1–4 year olds.

Motor vehicle crash deaths are most common among 5–19 year olds.

Child injury death rates decreased 11% from 2010 to 2019. However, rates increased among some groups.

↑50% Poisoning and drug overdose death rates among Hispanic children

↑37% Poisoning and drug overdose death rates among Black children

↑21% Suffocation death rates among Black children

↑9% Motor vehicle death rates among Black children

Focused prevention strategies can help prevent injuries and deaths.

Family engagement and support, parental monitoring, and school connectedness can reduce substance use.

Safe sleep strategies can reduce suffocation deaths among infants.

Proper use of car seats, booster seats, and seat belts can reduce motor vehicle crash injuries and deaths.

West BA, Rudd RA, Sauber-Schatz EK, Ballesteros MF. Unintentional injury deaths in children and youth, 2010–2019. Journal of Safety Research 2021;78:322-30. https://doi.org/10.1016/j.jsr.2021.07.001

**Fig. 1** Unintentional injury deaths in children and youth, 2010–2019. (Source: [1].) Image in public domain

The consequences of preventable injuries to children can result in lifelong disabilities, physical impairments, and psychosocial/emotional impacts.

Multiple organizations nationally advocate for child safety and injury prevention such as the Consumer Product Safety Commission, Safe Kids Worldwide, American Academy of Pediatrics, and, of course, CDCP [2–5]. The American College of Surgeons Committee on Trauma requires injury prevention and outreach activities to be conducted by trauma centers nationally. For trauma centers, injury prevention program activities should be reflective of their trauma data on mechanism of injury and targeted to local communities affected. Focusing on those injuries that occur most frequently is important, but a focus on those injuries that cause the most impact on the child, the most disability, to help reduce or eliminate those injuries is crucial as well. Pediatric surgeons can be critical in helping to get the message of injury prevention out to the public.

Adopting a plan for injury prevention education can make educational efforts easy. Focusing education based on seasonal activities that children and families engage in is very effective. In the spring, for example, children are getting outside again, so bike safety and helmet use are hot topics. Promoting correct helmet fit and use and a knowledge of bicycle traffic laws is important. Playground safety that includes supervision, safe equipment, and surfaces is key. Pedestrian safety focuses on "looking left, right, and left again", and crossing at cross walks not mid street or between cars. A community may need to evaluate their public safety plan to include signage and flashing lights in areas such as schools and parks where children spend more time. As summer approaches, water, pool, and boating safety become necessary to promote while continuing pedestrian and bike safety topics. Teaching children to swim is an essential parental responsibility. Utilizing lifejackets for all children on boats or near water is critical. Fireworks and severe weather education such as lightening safety are needed in the summer. In the winter, sledding, skiing, and ice-skating safety topics prevail along with winter travel safety. Home safety is important all year long with education on hot water temperatures, window safety guards, stair gates, crib and bed rails, safe storage of medications, cleaning solutions and other household items, and smoke and carbon monoxide detectors. Safe storage of firearms in the home is important for children of all ages. Another topic essential all year long is car seat safety education.

Since motor vehicle injuries are a leading cause of injury for children and teens, becoming familiar with some of the recommended car and booster seats for children and teen driving safety topics is key. Infants and toddlers should remain rear facing as long as possible, until the child outgrows their car seat's maximum height and weight guidelines. Rear-facing is the safest. For older toddlers and preschool size children, use a forward-facing seat with five-point harness for as long as possible, again up to the maximum size allowed by the seat. Booster seats for young school-age children should be used to ensure proper position of the lap/shoulder belt systems until the child is 4 ft 9 in tall, between ages 8 and 12, and above 80 pounds. Some general car seat safety is to never leave a child unattended in or around a vehicle and ensure the straps fit snugly and are correctly positioned and all children younger than age 13 are riding in the back seat. Involving a certified car seat technician to ensure child passenger safety by providing car seat education, checking car seat installation in a vehicle, and fitting children in car seats is beneficial. Many local police, State Police, and fire companies help to meet this need (Fig. 2).

Children are not ready to use a car seat belt restraint alone until they achieve a weight of 80 lbs, a height of 4 ft 9 in, and age 8–12 years old [6]. Car restraints are designed to fit adults, not children. Before allowing children to ride in a car with only a seat belt, they should be able to pass the five-step test (Table 1). Children should ride in the back seat until they are 13 years old.

Nationally, falls within the age range of 0–24 cause the highest number of injuries at nearly 29%, followed by "struck by or against" as the second leading mechanism of injury. Every day, approximately 8000 children are seen in US emergency

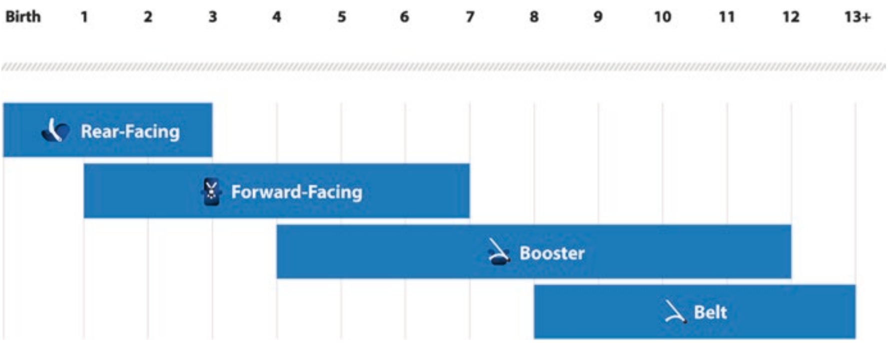

**Fig. 2** Recommended car seats based on your child's age and size. (Source: Car Seats and Booster Seats. National Highway Traffic Safety Administration. https://www.nhtsa.gov/equipment/car-seats-and-booster-seats, Accessed 26 Oct 2021) Image in public domain

**Table 1** The five-step test for children to ride in a vehicle using a seat belt without needing a car seat or booster seat

| | The five-step test for children to ride without a car seat or booster seat |
|---|---|
| 1 | Shoulder belt crosses over top of shoulder, not against neck |
| 2 | Child can sit with lower back against the vehicle seat back |
| 3 | The lap belt crosses the upper thighs against the hip bones, not the abdomen |
| 4 | Child can bend knees at end of seat while back is against seat back |
| 5 | Child can comfortably ride this way for entire ride |

Adapted from: Take the 5-Step Seat Belt Fit Test. Safe Ride 4kids. https://saferide4kids.com/blog/take-the-5-step-seat-belt-test/, Accessed 26 Oct 2021

rooms after having suffered a fall, which is approximately 2.8 million children yearly [7]. In the pediatric population, instituting fall prevention programs is challenging because of the varied ways children fall. Developmentally we expect young children to fall while learning to walk and run, but it's all the other ways children fall that become the challenge. Children fall off bicycles and monkey bars, fall on and off trampolines, riding a scooter, and fall out of shopping carts and off horses. All fall prevention education should be geared toward the developmental level of the child. Looking at your local trauma center data and the surrounding communities should direct your injury prevention program activities.

Injury prevention education should be presented both in person with interactive programs including education and activities. Safety messaging should be presented through the media and using social media outlets. During the COVID outbreak, many injury prevention messages had to be adapted for virtual presentations and through social media to reach the public.

When looking at injury prevention, we often focus on the most common causes of injury in the community being served, but it is essential also to look at contributing factors as well. Drugs, alcohol, behavioral and mental health issues, violence, poverty, and several other factors may be involved in poor childhood safety. An inability to afford cabinet or window locks can have a big impact on child safety during the toddler and preschool years. Acquiring grant funds to provide low cost or free resources to families is very beneficial. Several children's hospitals nationally host safety stores to provide low-cost items that underscore their safety and injury prevention efforts.

## Additional Notes

Safety is a team sport and not a competition between special interest groups. Use opportunities to work with partners with a similar focus such as police organizations to promote car seat safety and fire professionals for carbon monoxide and smoke detector use, and car dealerships and state departments of transportation for child passenger safety, to name a few. Larger organizations such as the Safe Kids Worldwide, the Injury Free Coalition for Kids, the American Trauma Society, and many other nonprofit groups form partnerships to promote safety messaging and program implementation. Any interaction with a child and family who has suffered a traumatic injury should be provided with safety education as part of their medical plan. Injury prevention will save far more lives than treatment afterward.

## Study Questions

1. What is the most common cause of death in children of all ages?
   a. Cancer
   b. Injury
   c. Pneumonia
   d. Heart disease

   Answer: (b.) Injury. For all age groups, child unintentional injury is the most common cause of death. More than 7000 children die each year as a result of unintentional injury. For infants, suffocation is the most common cause of death. Drowning is most common in children aged 1–4 years old. In children aged 5–19 years old, motor vehicle crash is the most common cause of death due to injury. Death due to injury is more common in male children and in both age groups: under 1 year old and 15–19 years old.

2. What is the proper vehicle restraint for a child who is 10 years old, weighs 85 lbs, and is 4 ft 2 in tall?
   a. Rear facing car seat
   b. Front facing car seat
   c. Booster seat and seat belt
   d. Seat belt alone

   Answer: (c.) Booster seat and seat belt alone. Children should use a restraint device that is appropriate to their age and size. Before a child graduates from a booster seat to sitting in a vehicle with a seat belt alone, they need to meet three criteria: aged 8–12 years old, weight 80 lbs. or greater, and height 4 ft 9 in or greater. Additionally, children should be able to pass the five-step test to be able to ride in a vehicle restrained by a seat belt alone: (1) shoulder belt crosses over shoulder, (2) lower back is against seat back, (3) lap belt crosses thighs and rests on hip bones, (4) knees bend at or past the seat edge, and (5) child can comfortably ride this way for entire trip.

## References

1. Injuries among Children and Teens. Centers for Disease Control and Prevention. https://www.cdc.gov/injury/features/child-injury/index.html. Accessed 26 Oct 2021.
2. Consumer Product Safety Commission. https://www.cpsc.gov/. Accessed 26 Oct 2021.
3. Safe Kids Worldwide. https://www.safekids.org/. Accessed 26 Oct 2021.
4. American Academy of Pediatrics. https://www.aap.org/. Accessed 26 Oct 2021.
5. Centers for Disease Control and Prevention. https://www.cdc.gov/. Accessed 26 Oct 2021.
6. Car Seats: Information for Families. Healthychildren.org, https://www.healthychildren.org/English/safety-prevention/on-the-go/Pages/Car-Safety-Seats-Information-for-Families.aspx. Accessed 26 Oct 2021.
7. Fall Prevention. Centers for Disease Control and Prevention. https://www.cdc.gov/safechild/falls/index.html. Accessed 26 Oct 2021.

# Non-accidental Trauma

**Carol Hanson**

**Key Points**

1. Child maltreatment can have lasting ill effects on a child development.
2. Identifying physical and nonverbal findings that may indicate a concern for child maltreatment.
3. Benefits of increased awareness in the medical community of subtle signs of child maltreatment as well as benefits of increase community education and awareness.

I. Abuse in children

  A. Non-accidental trauma (NAT) in children is considered an adverse childhood event (ACE). An ACE can have lasting impacts on a person's physical and mental health such as chronic health condition, depression, post-traumatic stress disorder (PTSD), substance abuse, or increased risk of suicide [1]. The Department of Health and Human Services uses the definition for abuse or neglect, at a minimum as:

    1. "Any recent act or failure to act on the part of a parent or caretaker which results in death, serious physical or emotional harm, sexual abuse or exploitation"
    2. "An act or failure to act which presents an imminent risk of serious harm" [2]

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B. State civil laws define the conduct, acts, and omissions that constitute abuse or neglect that must be reported to child protective agencies [3].
C. Sentinel injury: A previous injury reported in the medical history that was suspicious for abuse because child was too young to be able to cruise or explanation was implausible [4].
    1. A visible or detectable minor injury that is poorly explained and therefore suspicious for physical abuse.
    2. Bruises, intraoral injuries, certain fractures, pattern burns.
D. Sentinel injuries predict later abuse in children.
    1. Providers were aware of sentinel injuries in 41.9% of cases. Previous sentinel injuries are common in infants with severe abuse and rare in infants evaluated for abuse and not found to be abused [5].
    2. Concern of the ability of pediatric healthcare providers and social workers to recognize sentinel injuries in infants under 6 months old. Sentinel injuries not uniformly recognized as potential sign of child abuse.
E. Physical abuse is any physical injury inflicted upon a child with cruel and/or malicious intent. This can include striking, kicking, burning, biting, or any other physical action that results in a physical injury or physical impairment [3].
F. Emotional abuse or psychological abuse is when an adult uses behavior that negatively impacts a child's sense of self-worth in their emotional development. This may include demeaning, criticizing, and berating them in a manner that would humiliate and insult them [6].
G. Sexual abuse is defined as sexual activity with an adult, adolescent, or an older child. This includes touching or non-touching. Non-touching involves pornography and photography exposing genitals among other events [1].
H. Neglect. Child neglect is a behavior of caregivers where basic needs such as clothing, supervision, housing, or healthcare are withheld [3].
I. Physicians can miss fractures on prior examinations of abused children. In 20.9% of children with abusive fractures, there was at least one previous visit where abuse was missed. One fifth of children with abuse-related fractures are missed during initial medical visit [7].
J. Characteristic features of physical abuse: A child who is being abused may feel guilty, ashamed, or confused. He or she may be afraid to tell anyone about the abuse, especially if the abuser is a parent, other relatives, or family friend. It is vital to investigate for characteristic features [8].
    1. Withdrawal from friends or usual activities
    2. Changes in behavior such as aggression, anger, hostility, hyperactivity, or changes in school performance
    3. Depression, anxiety, unusual fears, or a sudden loss of self-confidence

4. An apparent lack of supervision
5. Frequent absences from school
6. Reluctance to leave school activities, as if he or she doesn't want to go home
7. Attempts at running away
8. Rebellious or defiant behavior
9. Self-harm or attempts at suicide
10. TEN-4 FACES P mnemonic of physical examination in NAT [9]
    a. A reminder of areas to check for injury when evaluating for abuse
    b. TEN
        i. Trunk
        ii. Ears
        iii. Neck
    c. 4
        i. In children 4 months old or younger, any bruises are suspicious for NAT because this is before most children are able to be mobile and cruise.
        ii. In children 4 years old or younger, bruises on the trunk, ears, or neck are suspicious for NAT.
    d. FACES
        i. Frenulum
        ii. Auricular area
        iii. Cheek
        iv. Eyes
        v. Sclera
    e. P
        i. Patterned bruising. Bruises in shapes and patterns of objects such as hand, a belt buckle, or parallel lines such as multiple strikes from a stick (Table 1)

II. Diagnostic studies in NAT

A. Bone scan

i. A bone scan examines all bones for any unidentified fractures.
ii. Multiple fractures, unreported fractures, and fractures of varying ages are suspicious for NAT.
iii. An additional bone scan should be performed 2 weeks after initial treatment. This amount of time allows for previously unidentified fractures to be identified through detection of callous formation.

**Table 1** Physical abuse assessment in non-accidental trauma

| Physical abuse assessment: signs of child abuse |
|---|
| A. Pre-verbal children (or nonverbal under age 18 years old) |
| 1. Bruising in children under 4 months old |
| 2. Seizure-like episodes at home |
| 3. Known intracranial bleed or abdominal bleed |
| 4. Known skeletal fractures |
| 5. Bruising to torso, ears, neck (TEN) area |
| 6. Traumatic injury warranting admission |
| 7. Acute life-threatening events (ALTE) |
| 8. Suspected deliberate toxic ingestion/obtained from another person |
| 9. Burns (including significant sunburn) |
| 10. Severely underweight |
| B. All children <18 years old |
| 1. Submersion injuries |
| 2. Strangulation injuries |
| 3. Suspected deliberate toxic ingestion/obtained by another person |
| 4. Injury out of proportion to history |

B. Head computed tomography (CT)

   i. Head CT is used to detect skull fractures and intracranial bleeding.
   ii. Abusive head trauma, such as when a baby is shaken, causes a characteristic pattern of bleeding.
   iii. Infants presenting with lethargy, dehydration, vomiting, and difficulty waking should be evaluated with head CT to determine if there has been abusive head trauma.

C. Spine magnetic resonance imaging (MRI)

   i. Spine MRI can detect ligamentous spinal injury and may detect subtle findings of edema in the spine musculature or spinal cord.
   ii. MRI spine identifies spinal subdural hemorrhage in most cases of abusive head trauma [10].

III. Consultations indicated in the evaluation of NAT in children

   A. Pediatric abuse specialist/pediatric hospitalist: Provide guidance for appropriate tests, diagnostic studies, photographic documentation, court/legal documentation, and testimony.
   B. Ophthalmologist: Provides evaluation and documentation of retinal hemorrhage, retinoschisis, and photographic documentation of retinal injuries.
   C. Case Management/Social Services: Provide guidance and referral to local community services.
   D. Child Protective Services/Children and Youth Services: Usually notified by Case Management or Social Services.
   E. Neurosurgery, orthopedic surgery, otolaryngology, oral maxillofacial surgery, etc.: Necessity depends on the injury complex.

IV. Notes of concern in the evaluation of NAT in children

    A. In one out of six skeletal surveys to detect NAT in children, the reading by a pediatric radiologist disagrees with the read from the general radiologist and results in significant change in the report [11].
    B. There is a high incidence of radiologic interpretation error in head computed tomography performed on abusive and non-abusive pediatric head trauma in children referred from a community hospital to a tertiary pediatric hospital [12].

V. Education on abuse and neglect: There is no standardized curriculum for medical students or residents on the topic of NAT. Many graduating physicians feel unprepared to address child abuse [13]. There is a need for improvement in the way physicians are trained to recognize and report abuse.

## Additional Notes

Awareness of child abuse prevalence and prevention has changed over the last half of the century. Recognition of child maltreatment has evolved from a personal concern to a community concern. These steps toward prevention of child maltreatment help to protect our children. Prevention also decreases the ongoing cycle of incidents seen through generations of the past. The benefits of NAT prevention are reduction of life-long physical and mental health disabilities, healthcare needs, treatment expenses, and lost opportunities in work and life goals.

## Study Questions

1. Child maltreatment and non-accidental trauma can be seen in which of the following populations:

    a. Low-income families
    b. Blue collar families
    c. White collar families
    d. Unrelated male in household
    e. Urban families
    f. Rural families
    g. Single parent families
    h. All of the above

    Answer: (h) All of the above. Abuse can affect children from any walk of life. It is important to avoid any bias on population identifiers to prevent missing any signs of child abuse by using screening tools and checklists in evaluating all injured children.

2. A parent brings a child to the emergency department saying that the child had some vomiting, has been irritable, and has been sleepy all day. He is having trouble getting child awake enough to eat. Should the provider be concerned?

    a. No, the child was probably had poor sleep last night and is only irritable.
    b. No, he may have a virus and needs intravenous fluid.
    c. Yes, irritability and prolonged sleepiness could be signs of something serious.

    Answer: (c) Yes, irritability and prolonged sleepiness could be signs of something serious. Young children are unable to give an account of what has happened to them. The signs of serious head trauma caused by abuse can mimic lethargy, dehydration, infection, and sepsis. It is important to consider NAT in all children who present with depression of mental status. Vomiting, irritability, lethargy, difficulty waking, and poor appetite can all be signs of head trauma.

3. Parents bring in an irritable 10-month-old child with a bump on his head after rolling off the couch. An evaluation reveals that the child has a skull fracture and an epidural bleed. Should child abuse be suspected?

    a. No – Kids roll around, fall off couches often, and injuries happen.
    b. Yes – It is child abuse. Parents need to be prosecuted.
    c. Unclear – A complete child abuse evaluation is indicated; this injury could be accidental but could also be NAT.

    Answer: (c) Unclear – A complete child abuse evaluation is indicated; this injury could be accidental but could also be NAT. It is important for providers to maintain a high index of suspicion of NAT. The history given may not be the actual events that occurred. Use of screening tools can reduce the chance that child abuse is missed. One possible indicator for screening is a child under the age of 1 year old injured within the home. This child should have a full evaluation for NAT including skeletal survey, retinal examination, and consultation of a pediatric abuse specialist.

## References

1. About Child Sexual Abuse. Prevent Child Abuse North Carolina, North Carolina Coalition for the Prevention of Child Sexual Abuse. https://www.preventchildabusenc.org/resource-hub/about-child-sexual-abuse/. Accessed 29 Oct 2021.
2. What is Child Abuse or Neglect? United States Department of Health and Human Services. https://www.hhs.gov/answers/programs-for-families-and-children/what-is-child-abuse/index.html. Accessed 28 Oct 2021.
3. Definitions of Child Abuse and Neglect. Child Welfare Information Gateway. U.S. Department of Health and Human Services, Children's Bureau, Washington, DC, 2019. https://www.childwelfare.gov/pubpdfs/define.pdf , Accessed 29 Oct 2021.
4. Petska HW, Sheets LK. Sentinel injuries: subtle findings of physical abuse. Pediatr Clin N Am. 2014;61(5):923–35.

5. Eismann EA, Shapiro RA, Thackeray J, Makoroff K, Bressler CJ, Kim GJ, Vavul-Roediger L, McPherson P, Izsak E, Spencer SP. Providers ability to identify sentinel injuries concerning for physical abuse in infants. Pediatr Emerg Care. 2021;37(5):e230–5.
6. Morin AL. What is emotional child abuse? Safety & First Aid. Verywell Family. About, Inc. Dotdash. https://www.verywellfamily.com/what-is-emotional-child-abuse-4157502 , Accessed 30 Oct 2021.
7. Ravichandiran N, Schuh S, Bejuk M, Al-Harthy N, Shouldice M, Au H, Boutis K. Delayed identification of pediatric abuse-related fractures. Pediatrics. 2010;125(1):60–6.
8. Child Abuse. Patient care and health information, diseases, and conditions. Mayo Foundation for Medical Education and Research. https://www.mayoclinic.org/diseases-conditions/child-abuse/symptoms-causes/syc-20370864. Accessed 30 Oct 2021.
9. Hinton C, Trop A. TEN-4 FACES P: a mnemonic to help you spot signs of child abuse. ACEP Now: Wiley; 2021. https://www.acepnow.com/article/ten-4-faces-p-a-mnemonic-to-help-you-spot-signs-of-child-abuse/. Accessed 31 Oct 2021.
10. Rabbitt AL, Kelly TG, Yan K, Zhang J, Bretl DA, Zuijano CV. Characteristics associated with spine injury on magnetic resonance imaging in children evaluated for abusive head trauma. Pediatr Radiol. 2020;50(1):83–97.
11. Kazmazyn B, Wanner MR, Marine MB, Tilmans L, Jennings SG, Hibbard RA. The added value of a second read by pediatric radiologists for outside skeletal surveys. Pediatr Radiol. 2019;49(2):203–309.
12. Kralik SF, Finke W, Wu IC, Hibbard RA, Hicks RA, Ho CY. Radiologic head CT interpretation errors in pediatric abusive and non-abusive head trauma patients. Pediatr Radiol. 2017;47(8):942–51.
13. Christian CW. Professional education in child abuse and neglect. Pediatrics. 2008;122(Suppl 1):S13–7.

## *Further Reading*

Schouten MCM, van Stel HF, Verheij TJM, Houben ML, Russel IMB, Nieuwenhuis EES, van de Putte EM. The value of a checklist for child abuse in out-of-hours primary care: to screen or not to screen. PLoS One. 2017;12(1):e0165641.

# Initial Trauma Assessment

**Dimitra M. Lotakis and Arthur Cooper**

**Key Points**

1) Mechanism of injury predicts pattern of injury, and pattern predicts response.
2) Differences in anatomy and physiology from adults result in responses to injury that are distinct from those seen in adults.
3) Most pediatric trauma is a disease of airway and breathing rather than bleeding and shock, because most major pediatric trauma is blunt trauma involving the head.

1. Pre-hospital care: Most pediatric trauma patients remain hemodynamically stable during initial triage and treatment. However, patients with polytrauma may suffer delayed and potentially rapid deterioration. Attention must also be directed to the fact that the age-specific anatomic and physiologic differences that characterize the several pediatric age groups (infant, toddler, preschooler, school-ager, adolescent) modify the response to traumatic injury typically observed in adults. It is under this lens which all patients must be assessed, maintaining a heightened level of awareness and examining the patient repeatedly.
    a. Basic life support

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i. Oxygen administration: High concentration humidified oxygen should be given to all patients based on the 2021 guidelines published under the title, "National Guideline for the Field Triage of Injured Patients", established by the American College of Surgeons Committee on Trauma and the US Centers for Disease Control and Prevention (Fig. 1). Delivery methods should be appropriately sized to patient. A non-rebreather mask, or nasal cannula, is the preferred method of administration, unless the patient requires assisted ventilation via self-inflating bag and mask. Airway adjuncts must also be considered. However, oropharyngeal airways may institute a gag reflex, vomiting, and aspiration in patients requiring bag-valve-mask ventilation who are not fully unresponsive. Nasopharyngeal airways are best avoided, owing to the abundance of easily damaged adenoidal tissue and the anterior location of the glottic inlet. Tracheal intubation via the oral route is indicated for those children in respiratory failure, decompensated shock, and/or traumatic coma if assisted ventilation via self-inflating bag and mask is ineffective; it is mandated for those in traumatic coma.

ii. Hemorrhage control: Most cases of external hemorrhage may be controlled by simple point pressure. Use of commercially manufactured arterial tourniquets to control hemorrhage secondary to extremity trauma is supported by consensus guidelines. Total ischemic time should be limited to the greatest extent possible. Although consensus guidelines for its use in children do not yet exist, use of topical hemostatic agent-impregnated gauze packing may also be employed, as no clear evidence exists to suggest deleterious effects.

iii. Spine stabilization: There are two main considerations in the preadolescent pediatric population. First, the relative craniofacial disproportion caused by the child's larger cranial dimension yet smaller jaw confers an alteration in the ratio of head to body size such that the child is predisposed to neck flexion when positioned supine. As a result, a 2.5 cm layer of padding should be placed below the patient extending from upper torso to pelvis in order to return cervical spine alignment to a neutral position. Second, functional residual capacity (FRC) can be greatly affected by strapping mechanisms used to secure a patient to the backboard. Special attention should be given to the placement of straps and any effects on respiratory function. Of note, children with trisomy 21 are prone to atlanto-axial instability, so spinal motion restriction is especially important in this subpopulation.

iv. Temperature maintenance: Due to the larger proportional body surface area, increased metabolic rate, and typically smaller amount of insulative tissue, the pediatric patient is uniquely susceptible to substantial changes in core body temperature and evaporative heat loss. Hypothermia, even relative, can affect the coagulation profile and increase morbidity. Care should be taken to reduce exposure time and maximize rewarming via

## National Guideline for the Field Triage of Injured Patients

### *RED CRITERIA*

### *High Risk for Serious Injury*

**Injury Patterns**

- Penetrating injuries to head, neck, torso, and proximal extremities
- Skull deformity, suspected skull fracture
- Suspected spinal injury with new motor or sensory loss
- Chest wall instability, deformity, or suspected flail chest
- Suspected pelvic fracture
- Suspected fracture of two or more proximal long bones
- Crushed, degloved, mangled, or pulseless extremity
- Amputation proximal to wrist or ankle
- Active bleeding requiring a tourniquet or wound packing with continuous pressure

**Mental Status & Vital Signs**

**All Patients**
- Unable to follow commands (motor GCS < 6)
- RR < 10 or > 29 breaths/min
- Respiratory distress or need for respiratory support
- Room-air pulse oximetry < 90%

**Age 0–9 years**
- SBP < 70 mm Hg + (2 x age in years)

**Age 10–64 years**
- SBP < 90 mmHg or
- HR > SBP

**Age ≥ 65 years**
- SBP < 110 mmHg or
- HR > SBP

***Patients meeting any one of the above RED criteria should be transported to the highest-level trauma center available within the geographic constraints of the regional trauma system***

### *YELLOW CRITERIA*

### *Moderate Risk for Serious Injury*

**Mechanism of Injury**

- High-Risk Auto Crash
  - Partial or complete ejection
  - Significant intrusion (including roof)
    - >12 inches occupant site OR
    - >18 inches any site OR
    - Need for extrication for entrapped patient
  - Death in passenger compartment
  - Child (age 0–9 years) unrestrained or in unsecured child safety seat
  - Vehicle telemetry data consistent with severe injury
- Rider separated from transport vehicle with significant impact (eg, motorcycle, ATV, horse, etc.)
- Pedestrian/bicycle rider thrown, run over, or with significant impact
- Fall from height > 10 feet (all ages)

**EMS Judgment**

**Consider risk factors, including:**

- Low-level falls in young children (age ≤ 5 years) or older adults (age ≥ 65 years) with significant head impact
- Anticoagulant use
- Suspicion of child abuse
- Special, high-resource healthcare needs
- Pregnancy > 20 weeks
- Burns in conjunction with trauma
- Children should be triaged preferentially to pediatric capable centers

**If concerned, take to a trauma center**

***Patients meeting any one of the YELLOW CRITERIA WHO DO NOT MEET RED CRITERIA should be preferentially transported to a trauma center, as available within the geographic constraints of the regional trauma system (need not be the highest-level trauma center)***

**Fig. 1** 2011 Guidelines for Field Triage of Injured Patients [1]

supplementing ambient environmental temperature, warming intravenous fluids when needed, and providing insulative coverings.

v. Fracture immobilization: Those fractures noted to be significantly displaced, unstable, or associated with active hemorrhage should be rapidly addressed in the field. Careful limb alignment, control of hemorrhage, and proper splinting are sufficient to facilitate safe transport.

    vi. Resuscitation tape: A length-based, color-coded resuscitation tape, such as the Broselow$^{TM}$ Tape (Armstrong Medical, Lincolnshire, IL), should be used to determine appropriate weight-based dosing and equipment sizes to facilitate rapid interventions. The tape should be laid out on the stretcher, preferably prior to the patient's arrival in the trauma bay.

b. Field triage: Current data support early triage and diversion to verified pediatric trauma centers. According to the American College of Surgeons Committee on Trauma National Guideline for the Field Triage of Injured Patients (Fig. 1), which have largely supplanted other approaches, initial assessment should focus on patients at high risk for serious injury (red criteria: injury patterns, mental status and vital signs), before those at moderate risk for serious injury (yellow criteria: injury mechanisms, EMS judgment). However, the following represent other verified scoring systems that may aid in field triage decision-making.

    i. Cognitive assessments: Glasgow Coma Score (GCS), which is included in the National Field Triage Criteria, provides a modality to rapidly evaluate cerebral function via best eye, verbal, and motor response assessments (Table 1). Similarly, the AVPU Score designates level of consciousness through one-letter designations (*A*lert, responsive to *V*oice, responsive to *P*ain, and *U*nresponsive). Unsatisfactory evaluations via these two systems remain one of the most sensitive and specific indicators of the need for specialized pediatric trauma care.
    ii. Pediatric trauma score (PTS): Rapid evaluation of body weight, airway status, systolic blood pressure, level of consciousness, and presence of fractures and cutaneous trauma yields a quantitative score created to aid in the assessment of multisystem trauma and need for specialty intervention. Originally designed as a "checklist" to aid prehospital providers in identifying the need for specialized pediatric trauma care, many emergency medical services (EMS) systems found the PTS too cumbersome for use when rapid triage decisions needed to be made.
    iii. Revised trauma score (RTS): This system utilizes GCS along with systolic blood pressure and respiratory rate to provide stratified assessments. Each metric is individually weighted, most significance being given to GCS. Similar to the PTS, the revised trauma score has significant limitations in its ability to reliably indicate need for specialty intervention. However, its heavy reliance on the GCS assessment does have its benefits, owing to the higher incidence of head trauma among injured children. It is important to note that the RTS is also limited in field use given its relative complexity vs. the National Field Triage Criteria.

**Table 1** Pediatric and adult Glasgow coma scale (GCS)

| Criteria | Infant | Child | Adult | Score |
|---|---|---|---|---|
| Eye opening | Spontaneous | Spontaneous | Spontaneous | 4 |
| | Verbal stimulus | Verbal stimulus | Verbal stimulus | 3 |
| | To pressure | To pressure | To pressure | 2 |
| | None | None | None | 1 |
| (Criterion total) | | | | (1–4) |
| Verbal response | Coos, babbles | Oriented, appropriate | Oriented | 5 |
| | Irritable cries, consolable | Confused | Confused | 4 |
| | Inconsolable, moans | Inappropriate words | Inappropriate words | 3 |
| | Inconsolable, agitated | Incomprehensible | Incomprehensible | 2 |
| | None | None | None | 1 |
| (Criterion total) | | | | (1–5) |
| Motor response | Spontaneous, purposeful | Obeys command | Obeys command | 6 |
| | Withdraw to touch | Localizes to pressure | Localizes to pressure | 5 |
| | Withdraw to pressure | Withdraw to pressure | Withdraws to pressure | 4 |
| | Flexion to pressure (decorticate posture) | Flexion to pressure (decorticate posture) | Flexion to pressure (decorticate posture) | 3 |
| | Extension to pressure (decerebrate posture) | Extension to pressure (decerebrate posture) | Extension to pressure (decerebrate posture) | 2 |
| | None | None | None | 1 |
| (Criterion total) | | | | (1–6) |
| Total GCS | | | | 3–15 |

2. In-hospital care
    a. Considerations for injury patterns: Several characteristic injury patterns have been observed in pediatric patients based on common childhood injury mechanisms. While these mechanisms do not guarantee the presence of these specific injuries, an increased level of suspicion should be maintained during both initial field triage and primary assessment.
        i. Motor vehicle occupant: Unrestrained passengers most commonly suffer injuries of the head, face, and neck. Restrained passengers may present with injuries to the cervical spine, Chance fractures of the lower thoracic and lumbar spine, and/or disruption or hematoma of the small intestine.
        ii. Pedestrian struck: If the vehicle is traveling at low speed, patients generally suffer only lower extremity injuries. However, if traveling at high speed, patients are more likely to experience polytrauma including head, neck, and torso injuries as well as lower extremity injuries.

   iii. Bicyclist: Those patients who cycle without helmets often sustain traumatic brain injuries and head and neck lacerations as well as upper extremity fractures. As expected, those with helmets are usually seen with injuries limited to the upper extremities. Special consideration should be given to any patient with a history of impact upon the handlebar, as this mechanism indicates higher propensity for intra-abdominal trauma.
   iv. Falls: Fall injury patterns are stratified by height of fall in relation to patient height. Ground-level falls typically result in isolated upper extremity fractures. Mid-level heights produce injuries of head, neck, and upper or lower extremities based on area of impact. Falls from height produce polytrauma with similar patterns as mid-level falls as well as damage to intra-thoracic and intra-abdominal organs.

b. Primary survey: As with their adult counterparts, initial assessment serves to identify life-threatening conditions in order to deliver treatment aimed not only toward survival but to diminishing the consequences of secondary injury as well.

   i. Airway: Airway patency and adequacy of tissue oxygenation should be addressed immediately upon arrival in the trauma bay and continually reassessed throughout the duration of the trauma evaluation. Those patients identified with compromised airways or inability to protect their airways due to level of consciousness or injury pattern should be orotracheally intubated. Supplemental oxygen should be administered to any patient meeting National Field Triage Criteria and in any patient exhibiting signs of respiratory insufficiency, circulatory instability, or neurologic injury. Specific considerations for the anatomy of the pediatric airway include its narrower diameter at all levels; the anterior, cephalad, funnel-shaped larynx; and the short trachea.
   ii. Breathing: As a clinical entity, pediatric trauma is primarily a disease of airway and breathing rather than bleeding and shock, because most major pediatric trauma is blunt trauma involving the head. Thorough evaluation, early detection, and prompt management of any abnormalities are critical to favorable outcomes. Auscultatory examination as well as visual inspection and palpation should be completed to identify significant injuries such as tension pneumothorax. It is important, however, to ensure the adequacy of gas exchange. Inadequacy of ventilation and oxygenation is the most common cause of traumatic cardiac arrest in the pediatric population. Waveform capnography, pulse oximetry, and arterial blood gas analysis will assist in monitoring for potential abnormalities. For infants and children, normal spontaneous tidal volumes are 6–8 mL/kg. It is not unusual, however, for volumes as high as 7–10 mL/kg to be required during assisted ventilation.

iii. Circulation: The six components of initial circulatory assessment include early identification and control of significant external hemorrhage; palpation of central and peripheral pulses; appraisal of capillary refill time; evaluation of skin color, temperature, and moisture; measurement of blood pressure; and reassessment of level of consciousness. It is these metrics which provide the most robust data to allow early diagnosis and management of circulatory derangement. However, owing to their greater physiologic reserve, it is important to note that pediatric trauma patients can sustain up to 25–30% loss of total blood volume before they manifest hemodynamic changes consistent with hypotensive shock. Heralding signs of impending compromise include weakening of peripheral pulses, prolongation of capillary refill, and narrowed pulse pressure, in addition to decreased level of consciousness. Rough estimation of the lower limit of normal systolic blood pressure can be made by multiplying the patient's age by 2 and adding 70 [(2 × Age) + 70].

iv. Disability: It is at this time that reassessment of neurologic status is key, specifically noting any signs of impending herniation or neurologic injury such as changes or asymmetry in the pupillary exam, bulging fontanelles (in infancy), and changes in level of consciousness or unresponsiveness. Prevention of secondary brain injury is paramount in cases with any indications of cerebral trauma. This is accomplished by maintaining cerebral perfusion pressure while controlling intracranial pressure and ensuring adequate tissue oxygenation. The Glasgow Coma Scale (GCS) is used in pediatric patients, though modifications have been suggested for infants and preverbal children (Table 1).

v. Exposure: Full exposure of all body surfaces, including upper and lower back, bilateral axillae, and perineum, are important to identification of injuries. Prompt provision of warmed blankets and maintenance of ambient temperature are critical, as previously discussed.

vi. Life-threatening injuries: The following injuries must be rapidly identified during the primary survey as rapid correction requires immediate attention. Some of the injuries may require intervention while completing the primary survey.

    a) Airway obstruction
    b) Respiratory failure
    c) Tension pneumothorax
    d) Sucking chest wound
    e) Flail chest
    f) Ongoing hemorrhage
    g) Pericardial tamponade
    h) Shock

vii. Vital signs and monitoring: Clinical and electronic monitoring of vital signs during the initial phases of resuscitation is essential to the early recognition of physiologic deterioration. Heart rate, respiratory rate, blood pressure using an appropriately sized cuff, pupillary reactivity, size, and symmetry, as well as Glasgow Coma Scale (GCS) score, should be recorded on a paper- or computer-based Trauma Flow Sheet every 3–5 min until stability can be established. It is helpful to post age-specific vital signs on a wall chart in the trauma bay (Table 2).

viii. Family presence: Several studies in recent years have demonstrated the potential value of family presence during resuscitation, so long as proper order can be maintained. Family presence allows parents and/or guardians to be with their child during these difficult moments, enhancing their sense of parental responsibility to protect their child while facilitating their recognition that everything is being done to restore their child's health in as timely and professional manner as possible.

c. Secondary survey: This portion of the initial assessment is comprised of a complete head to toe examination aimed at identifying injuries responsible for the physiologic derangements identified during the primary survey or not found during the primary survey. Providers must also elicit the salient "SAMPLE" history from patient, parent, or caregiver, including *S*ymptoms ("What hurts?"), *A*llergies, *M*edications, *P*ast medical history, *L*ast meal, and the *E*xposures/*E*vents ("What happened?") associated with the observed injuries.

i. Head: Dedicated exam of fontanelles in the infant population is critical to identifying signs of increased intracranial pressure, indicating intracranial hemorrhage or cerebral edema. Soft tissue and bony injuries of the head and face are common across multiple mechanism types. Re-evaluation of pupillary symmetry and response in addition to otoscopic

**Table 2** Normal vital signs for pediatric patients based on age and weight

| Age | Weight (kg) | Heart rate | Systolic BP | Respiratory rate |
|---|---|---|---|---|
| Preterm | <2.5 | 120–180 | 40–60 | 55–65 |
| Neonate | 3 | 90–170 | 50–90 | 40–60 |
| 1 month | 4 | 110–180 | 66–104 | 30–50 |
| 6 months | 7 | 110–180 | 72–110 | 25–35 |
| 1 year | 10 | 80–160 | 72–110 | 20–30 |
| 2–3 years | 12 | 90–150 | 74–124 | 18–30 |
| 4–5 years | 17 kg | 65–135 | 79–119 | 18–30 |
| 6–8 years | 24 kg | 70–115 | 77–118 | 18–24 |
| 8–10 years | 28 kg | 65–110 | 80–118 | 18–22 |
| 10–12 years | 35 kg | 55–110 | 84–131 | 14–22 |
| 13–14 years | 50 kg | 55–110 | 92–140 | 12–20 |
| 15–18 years | 70 kg | 55–110 | 100–140 | 12–20 |

visualization of tympanic membranes should also be completed. Specific inspection for drainage of fluid via nares, any midface instability, and periorbital and posterior auricular ecchymosis is critical when considering the possibility of craniofacial and/or basilar skull fractures.

ii. Neck: A certain level of skeptical awareness should be maintained with respect to injuries to the neck. Despite the greater angular momentum forces associated with their larger heads, their increased ligamentous elasticity, and their horizontally aligned vertebral facet joints, children are generally less susceptible to cervical spine and spinal cord injury. However, vertebral and spinal cord injuries do occur, and the latter often without radiographic confirmation, a condition known as Spinal Cord Injury With Out Radiographic Abnormality (SCIWORA). In the event that examination findings raise concern for cervical spine or spinal cord injury, in-line spinal motion restriction should be maintained with a semi-rigid extrication collar.

iii. Chest: Careful inspection, palpation, auscultation, and percussion should be carried out in order to identify soft tissue, bony, and pulmonary injury. If chest trauma is identified, it is important to complete a thorough exam of all other body regions, as up to 65% of such patients will have injuries elsewhere. Any external signs of trauma such as ecchymosis, subcutaneous emphysema, or significant tenderness with palpation should prompt further investigation for associated injuries.

iv. Abdomen: Adequate and accurate abdominal examination can be challenging in the pediatric population. One must ensure the child is as calm and quiet as achievable. This will allow for reduction in voluntary guarding and improve evaluation during palpation. In young children, elevating the legs with flexion at the hip may help relax abdominal muscles to improve sensitivity of palpation. This, however, should only be performed in patients whom lower extremity and thoracic spinal trauma are not suspected. Gentle pressure should be initially applied with gradual increase in depth in order to ease into the exam and moderate pain. Placement of a gastric tube and/or bladder catheter for decompression may be necessary to facilitate examination. Ecchymosis of the lower abdomen following motor vehicle collision (seat-belt sign) and ecchymosis of the upper abdomen from impact with solid objects such as handlebars are warning signs for concomitant intra-abdominal injury, specifically injuries to upper abdominal viscera.

v. Pelvis: The pelvic inlet is significantly shallower than during adulthood. Though fractures of the pelvis occur at lower rates than in adult trauma patients, bladder rupture is known to be more prominent in children. Visual inspection and palpation for stability are key during this portion of the secondary survey.

vi. Extremities: Prior to puberty and late adolescence, the periosteum of cortical bone remains porous, giving it an increased level of elasticity and flexibility. Further consideration must be given to the presence of open growth plates. These anatomic and physiologic differences lead to higher rates of incomplete fractures. As previously discussed, many mechanism types are associated with both upper and lower extremity trauma resulting in bony and soft tissue injuries. As such, any anatomic deformity, swelling, or ecchymosis should be investigated with appropriate imaging, recognizing that plain radiographs may not demonstrate fractures, when present, until 7–10 days post-injury.

d. Diagnostic imaging: Selective and judicious use of radiographic imaging is important as radiation exposure is not without risk. Every effort should therefore be made to limit radiographic imaging only to those body areas for which it is needed to guide management.

   i. Plain radiography: Anteroposterior chest radiographs are useful in those patients who suffered significant blunt force or penetrating trauma to the chest. Sensitivity and specificity are favorable in pediatric patients. Pelvic imaging is of lesser utility, given that pelvic fractures are uncommon and often not accurately identified. In a cooperative patient with a concerning mechanism and suggestive examination findings, cervical spine imaging may be considered if injury cannot be excluded via clinical examination (Table 3).

   ii. Ultrasound: While *F*ocused *A*ssessment by *S*onography in *T*rauma (FAST) examination has shown much promise in the adult trauma population, its application is less well established in pediatric practice. Its purpose is early recognition of internal bleeding (pooling of blood in the pericardial sac, Morison's pouch, the splenic fossa, and the cul-de-sac), while in recent years, it has been *E*xtended (E-FAST) to identify pneumothorax and/or hemothorax (collections of blood and/or air in the hemithoraces). Its lesser sensitivity and specificity, hence inability to definitively exclude the presence of intra-abdominal injury, mean it may not add significantly to the evaluation of a child with abdominal trauma. Those

**Table 3** Adapted from Pediatric Emergency Care Applied Research Network (PECARN) guidelines for cervical spine imaging in children <16 years of age [4]

| |
|---|
| Altered mental status |
| Focal neurologic findings |
| Neck pain |
| Torticollis |
| Substantial torso injury |
| Conditions predisposing to cervical spine injury |
| Diving |
| High-risk motor vehicle crash |

Having one (1) or more of the above factors warrants cervical spine imaging patients who are hemodynamically stable with concerning mechanism, examination, and laboratory findings should likely undergo CT imaging. The presence of hemorrhagic shock necessitates exploration in the operating room without delay.

iii. Computed tomography: When determining need for CT imaging, benefit of diagnosis must outweigh risk of radiation exposure. The Pediatric Emergency Care Applied Research Network (PECARN) has established guidelines that define those circumstances in which head and abdominal CT can be safely deferred. In general, any patient suffering more than momentary loss of consciousness or with abnormal GCS score should undergo head CT without contrast given the concern for intracranial pathology and the potential need for immediate intervention should pathology be identified. Those patients with normal mental status, no loss of consciousness, relatively benign mechanism, and injury patterns do not require imaging of the head (Fig. 2). In the case of cervical spine and thoracic injury, the added benefit of CT imaging is often limited while adding the burden of additional radiation exposure. However, abdominal CT with intravenous, and if feasible, oral contrast remains the "gold standard" for evaluation of potential intra-abdominal injuries (Fig. 3).

e. Laboratory studies: As a result of their relatively limited sensitivity and specificity, routine laboratory studies need not be performed in pediatric trauma patients. Selective use, however, has been shown to improve diagnosis and guide management. Patients with compromised respiratory status or neurologic injury benefit from arterial blood gas analysis, as adequate ventilation, oxygenation, and perfusion are key to minimizing secondary injury. If examination findings concerning for intra-abdominal trauma are observed, such as the seat-belt sign or epigastric ecchymosis, consideration should be given to hepatic function testing along with amylase and lipase. Serial examination of hemoglobin and hematocrit is far more useful than single-point testing in patients whom hemorrhage is a concern. Urinalysis is required to identify the need for renal imaging, which is indicated if more than 50 red blood cells (RBCs) are observed per high power field (HPF).

f. Vascular access: In any patient with hemodynamic instability or signs of neurologic injury, vascular access is critical to optimize outcome. Many routes are available to accomplish this goal. Should multiple attempts be made via one method without success, transition to other techniques must be undertaken rapidly in order to obtain definitive access.

    i. Peripheral intravenous line: As with adults, this is the favored method of access, since their short length and relatively large caliber ensure minimal resistance to flow during resuscitation. The antecubital fossa and saphenous veins at the level of the ankle are preferred. Generally, two attempts are made. If unable to obtain, then alternative methods are sought. Catheter sizing is dependent upon age and weight.

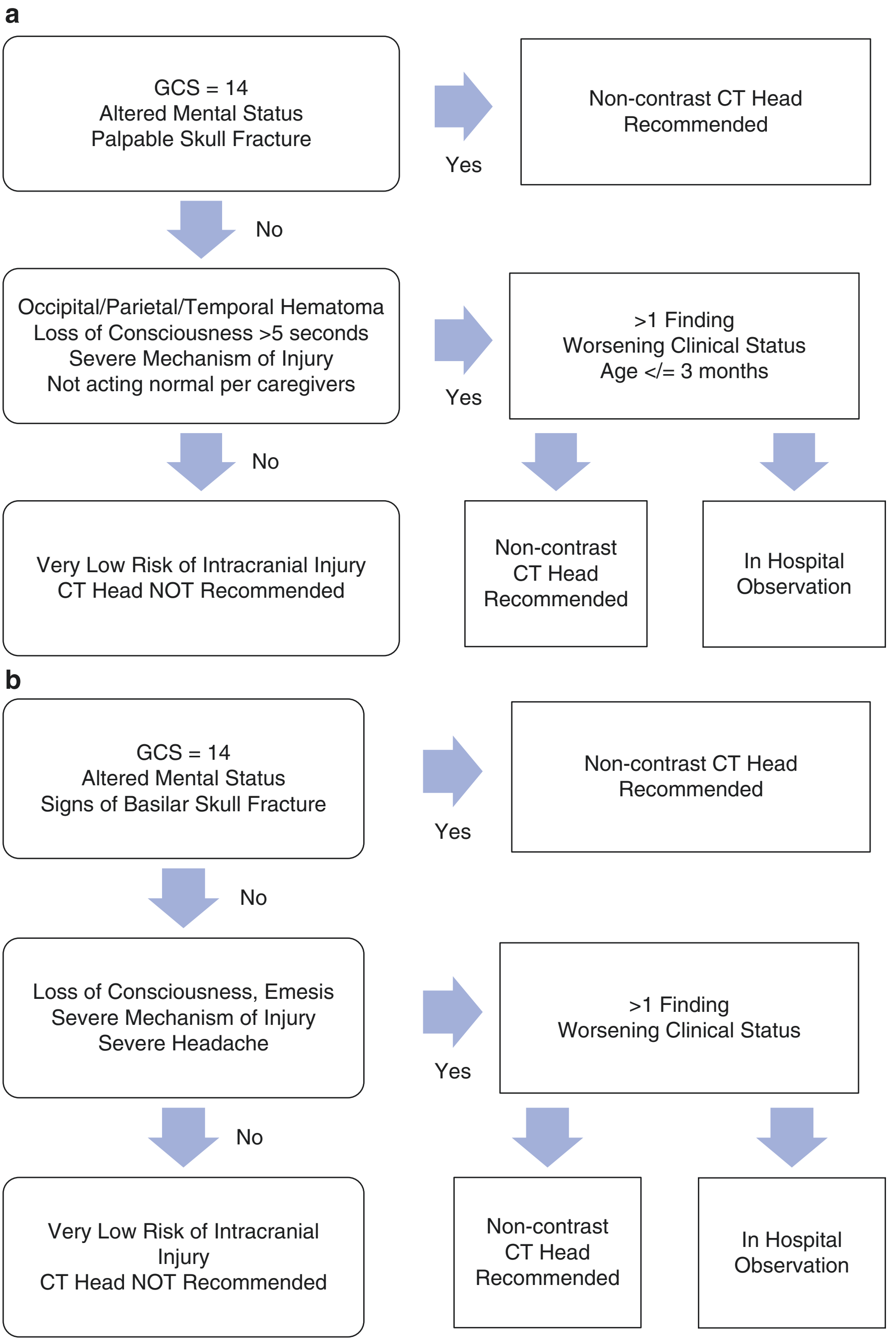
a
GCS = 14
Altered Mental Status
Palpable Skull Fracture
Yes
Non-contrast CT Head Recommended
No
Occipital/Parietal/Temporal Hematoma
Loss of Consciousness >5 seconds
Severe Mechanism of Injury
Not acting normal per caregivers
Yes
>1 Finding
Worsening Clinical Status
Age </= 3 months
No
Very Low Risk of Intracranial Injury
CT Head NOT Recommended
Non-contrast CT Head Recommended
In Hospital Observation
b
GCS = 14
Altered Mental Status
Signs of Basilar Skull Fracture
Yes
Non-contrast CT Head Recommended
No
Loss of Consciousness, Emesis
Severe Mechanism of Injury
Severe Headache
Yes
>1 Finding
Worsening Clinical Status
No
Very Low Risk of Intracranial Injury
CT Head NOT Recommended
Non-contrast CT Head Recommended
In Hospital Observation

**Fig. 2** Adapted from Pediatric Applied Care Research Network (PECARN) guidelines for head CT. (**a**) Younger than 2 years of age. (**b**) 2 years of age and older. Reused with permission from ref. [2]

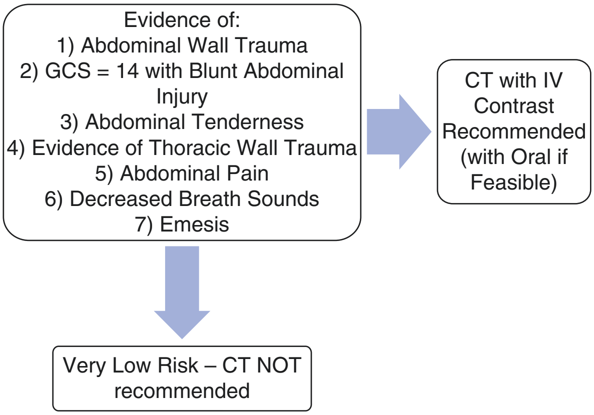
Evidence of:
1) Abdominal Wall Trauma
2) GCS = 14 with Blunt Abdominal Injury
3) Abdominal Tenderness
4) Evidence of Thoracic Wall Trauma
5) Abdominal Pain
6) Decreased Breath Sounds
7) Emesis
CT with IV Contrast Recommended (with Oral if Feasible)
Very Low Risk – CT NOT recommended

**Fig. 3** Adapted from Pediatric Applied Care Research Network (PECARN) guidelines for abdominal CT. Reused with permission from ref. [3]

ii. Intraosseous: This is an excellent adjunct should peripheral access prove challenging. Anterior tibial, distal femur, iliac crest, and distal tibia are all potential access sites based on age and weight, the former being the strongly preferred site, unless a fracture is present in the same lower extremity.

iii. Percutaneous femoral: Should peripheral and intraosseous attempts fail, percutaneous access of the femoral vein may be attempted. Advantages to this location include the ability to continue cardiopulmonary resuscitative efforts if previously initiated. Ultrasound guidance should be used when feasible, but placement guided by appropriate anatomic landmarks is generally safe. Seldinger technique should always be employed.

g. Volume resuscitation: Once secure vascular access has been obtained, intravenous fluid should be administered at an hourly maintenance rate according to the Holliday-Segar formula (100 mL/kg for the first 10 kg of body weight, plus 50 mL/kg for the second 10 kg of body weight, plus 20 mL/kg for every additional kilogram of body weight, all divided by 24). However, if there is any indication of circulatory or hemodynamic instability, volume resuscitation is indicated.

   i. Calculating volume: First define the patient's weight as accurately as possible. If unable to obtain quantitative weight via specialized gurney or stretcher, tools for assessment include eliciting history from caregiver, estimation via resuscitation tape, or simple calculation. A rough estimate can be obtained by multiplying the age by 2 and adding 10. Subsequent volume administered is based on a specified mL/kg equivalent. This also applies to various medications.

   ii. Choice of fluid: In patients presenting with mild hypotension not associated with active hemorrhage, 20–40 mL/kg of warmed lactated Ringer's solution may be delivered with observation for responsiveness. For patients with evidence of significant or ongoing hemorrhage, early use of "balanced resuscitation" utilizing 10–20 mL/kg packed red blood cells, 10–20 mL/kg of plasma frozen within 24 h of phlebotomy (PF24) in lieu of fresh frozen plasma, and single donor platelets should be considered, although the benefits of balanced resuscitation vs. traditional component therapy have not been conclusively established.

h. Gastric tube and bladder catheter: Placement of decompressive tubes becomes particularly useful if the abdominal exam is limited by distension. Intubated patients should have both a gastric tube and a bladder catheter placed unless contraindications are present. Similar to adult trauma, clinical concern for craniofacial fractures represents contraindications to gastric tube placement via the nasal route, while the possibility of urethral injury should prompt retrograde urethrogram before bladder catheterization is attempted. If such injury is found, suprapubic catheterization by qualified physician should be considered.
i. Vital signs and monitoring: As in adult trauma, multiple markers of resuscitation should be followed in the pediatric trauma patient, in addition to vital signs, which as previously noted are dependent on age and weight (Table 2).
    i. Urine output: Infants (2 mL/kg/h), younger children (1.5 mL/kg/h), older children (1 mL/kg/h)
    ii. ABG/VBG: Obtain as needed based on injury mechanism and method of ventilation

## Additional Notes

### *Historical Perspective*

Traumatic injuries are synonymous with the dawn of humankind. Documentation extending as far back as the early Egyptians has demonstrated the treatment of traumatic injury. Napoleon's chief military surgeon, Baron Dominique Jean Larrey, was the first to apply principles of triage, treatment, and transport from the battlefield in the late eighteenth century. Nineteenth-century physicians, including William Beaumont, Samuel D. Gross, and Nicholas Senn, worked tirelessly to document their experiences with victims of trauma as well as experiments with animal models in trauma. The greatest advances in the early twentieth century included development of the half-ring traction splint for reduction and immobilization of femur fractures by Hugh Owen Thomas and the use of occlusive dressings and chest decompression for sucking chest wounds by Evarts Ambrose Graham.

Despite these efforts, the advent of modern trauma management did not occur until the mid to late twentieth century. Following the World Wars, interest peaked regarding further classification and treatment of traumatic injuries in order to improve outcomes, leading to marked improvements in triage, treatment, and transportation during the Korean and Vietnam conflicts. However, it was not until the 1960s that major advances in both prehospital emergency medical services and cardiopulmonary and volume resuscitation led to development of dedicated trauma centers and organized trauma systems. In 1962, the first Pediatric Trauma Center was developed at Kings County Hospital in Brooklyn, New York by Dr. Peter Kottmeier. Subsequent research endeavors have shown improved quality and outcomes in patients receiving care at specialized centers.

As care for pediatric trauma victims became more centralized, research efforts expanded, and as a result, true understanding of mechanism of trauma and the resultant injury patterns began to fully form. It was the increased understanding of the interplay among physiology, anatomy, and mechanism that helped delineate pediatric trauma as a unique field from the care of adult patients. Arguably one of the most important advances in care of the pediatric trauma patient came with the publication of Dr. Sigmund Ein's 1978 paper, "Nonoperative management of traumatized spleen in children: How and Why," based on the experience at the Hospital for Sick Children in Toronto, Ontario. Though the initial response was one of skepticism, further evaluation and investigation led to our current nonoperative management protocols of multiple types of solid organ injury including spleen, liver, and kidney.

### *Future Directions*

The greatest challenge in the management of pediatric trauma remains its prevention, as up to 80% of childhood injuries could be prevented by the application of well-established injury prevention methods already at our disposal. Other challenges include the approach to blood replacement in initial management, including application of massive transfusion protocols, use of balanced resuscitation techniques, and the role of tranexamic acid, as well as viscoelastic monitoring of trauma-related coagulopathy, the proper role damage control strategies, and the perennial search for improved techniques of cerebral resuscitation.

## Study Questions

1) Question 1: A 10-year-old boy is brought in by EMS after being struck by a car while riding his bicycle. Upon arrival his GCS score was 12, his blood pressure 81/52 mm Hg, his heart rate 145 beats/min, his respiratory rate 40 breaths/min, and his oxygen saturation 95%. He has a prominent temporal-parietal hematoma. During the primary survey, his oxygen saturation begins to decline to

80%. After administration of 12L oxygen via non-rebreather mask, his oxygen saturation improves to 92%. At this time, the primary survey is continued, but his GCS score has declined to 10. The most important next step in management is:

a. Complete primary survey and obtain non-contrast head CT.
b. Complete primary survey, and consult neurosurgery for further instructions.
c. Complete primary and secondary surveys, reassess GCS score, and determine need for head CT.
d. Perform endotracheal intubation before continuing with primary survey.

Answer to Question 1: (d.) Given the mechanism of injury, notable scalp hematoma, initial instability, and neurologic deterioration, as indicated by the decline in GCS score by 2 points, the next most important step is to secure the airway. This patient is high risk for intracranial injury which may significantly compromise respiratory function and subsequently result in hypoxemia leading to secondary injury.

2) Question 2: A 12-year-old girl is brought in by EMS following a high-speed MVC in which she was a restrained rear seat passenger. Upon arrival her blood pressure is 92/76 mm Hg, her heart rate of 134 beats/min, and her oxygen saturation 95%. Primary survey is notable for a prominent transverse seatbelt sign and abdominal tenderness. She is treated with a 20 mL/kg IV bolus of lactated Ringer's solution with good response. After completion of the primary and secondary surveys, which were otherwise intact, she begins to report significant abdominal pain. The appropriate next step in management would be:

a. Proceed directly to the operating room for an exploratory laparotomy.
b. Obtain an abdominal CT with IV contrast.
c. Complete a FAST exam; discharge home if normal.
d. No additional workup; discharge home after tertiary survey complete.

Answer to Question 2: (b.) Given this patient's significant evidence of abdominal wall trauma (seat belt sign, pain, and tenderness), she would meet criteria for increased risk of intra-abdominal injury based on the PECARN guidelines. In this case, abdominal CT with IV contrast, and ideally PO if feasible, is recommended.

3) Question 3: A 6-year-old is struck by a car while playing in the street just in front of his driveway. On arrival he is moving all his extremities spontaneously, opens eyes to verbal stimuli, and is crying that his head hurts. His mother reports he was unresponsive initially, then recovered, and immediately vomited. Given an otherwise normal primary and secondary survey, what additional care strategy would you employ?

a. Do nothing; discharge home with Tylenol for headaches.
b. Admit to inpatient unit for observation with Tylenol PRN for headaches.
c. Obtain non-contrast head CT.
d. Admit to PICU for inpatient observation with neurologic checks every hour.

Answer to Question 3: (c.) Based on the PECARN guidelines, given this child is greater than 2 years old with multiple symptoms including loss of consciousness, emesis, and headaches, the recommendation would be to screen for intracranial injury with non-contrast head CT due to an increased risk of clinically important brain injury.

## References

1. Newgard CD, Fischer PE, Gestring M, Michaels HN, Jurkovich GJ, Lerner EB, Fallat ME, Delbridge TR, Brown JB, Bulger EM, For the 2021 National Expert Panel on Field Triage. National guideline for the field triage of injured patients: recommendations of the national expert panel on field triage, 2021. J Trauma Acute Care Surg. 2022 (epub ahead of print).
2. Kuppermann N, Holmes JF, Dayan PS, Hoyle JD, Atabaki SM, Holubkov R, Nadel FM, Monroe D, Stanley RM, Borgialli DA, Badawy MK, Schunk JE, Quayle KS, Mahajan P, Lichenstein R, Lillis KA, Tunik MG, Jacobs ES, Callahan JM, Gorelick MH, Glass TF, Lee LK, Bachman MC, Cooper A, Powell EC, Gerardi MJ, Melville KA, Muizelaar JP, Wisner DH, Zuspan SJ, Dean JM, Wootton-Gorges SL, for the Pediatric Emergency Care Applied Research Network (PECARN). Identification of children at very low risk of clinically-important brain injuries after head trauma: a prospective cohort study. Lancet. 2009;374:1160–70.
3. Holmes JF, Lillis K, Monroe D, Borgialli D, Kerrey BT, Mahajan P, Adelgais K, Ellison AM, Yen K, Atabaki S, Menaker K, Bonsu B, Quayle KS, Garcia M, Rogers A, Blumberg S, Lee L, Tunik M, Kooistra J, Kwok M, Cook LJ, Dean JM, Sokolove PE, Wisner DH, Ehrlich P, Cooper A, Dayan PS, Wooton-Gorges S, Kuppermann N. Pediatric Emergency Care Applied Research Network (PECARN). Identifying children at very low risk of clinically important blunt abdominal injuries. Ann Emerg Med. 2013;62(2):107–16.
4. Leonard JC, Kuppermann N, Olsen C, Babcock-Cimpello L, Brown K, Mahajan P, Adelgais KM, Anders J, Borgialli D, Donoghue A, Hoyle JD, Kim E, Leonard JR, Lillis KA, Nigrovic LE, Powell EC, Rebella G, Reeves SD, Rogers AJ, Stankovic C, Teshome G, Jaffe DM. for the Pediatric Emergency Care Applied Research Network. Factors associated with cervical spine Injury in children after blunt trauma. Ann Emerg Med. 2011;58:145–55.

## *Further Reading*

Anonymous. Pediatric trauma. In: American College of Surgeons Committee on Trauma. ATLS®: Advanced Trauma Life Support® Student Course Manual. 10th ed. Chicago, IL: American College of Surgeons; 2018. p. 186–211.

Cooper A. Early assessment and management of trauma. In: Holcomb GW, Murphy J, St. Peter SD, editors. Holcomb and Ashcraft's pediatric surgery. 7th ed. Philadelphia: Elsevier; 2019. p. 211–23.

# Pathophysiology/Pathobiology of Brain Injury

**Nelson Sofoluke, George Jallo, and Nir Shimony**

**Key Points**

1) Traumatic brain injury is one of the leading causes of death in the pediatric population.
2) Significant effort and research have been made to improve the diagnosis, monitoring, and treatment of pediatric patients with traumatic brain injuries.
3) The most important aspect is to maintain cerebral perfusion pressure and avoid secondary brain injury.

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1. Definitions
    a. Pediatric/Infant Glasgow Coma Scale (GCS) (Tables 1 and 2)
    b. Traumatic Brain Injury (TBI)
        i. TBI is the leading cause of pediatric traumatic death, accounting for approximately 37,000 admissions and 2500 deaths per year in the United States (US).
        ii. TBI is classified as: (see Table 3)
            1. Mild
                a. An alteration or loss of consciousness as a result of non-penetrating traumatic injury to the brain that lasts ≤30 min with or without post-traumatic amnesia [1]; GCS 13–15
                b. Findings include a vacant stare, delayed verbal or motor responses, difficulty focusing, memory deficits, and irritability
            2. Moderate
                a. An alteration or loss of consciousness that lasts >30 min [1]; GCS 9–12
            3. Severe
                a. An alteration or loss of consciousness >30 min that can progress to coma [1]; GCS 3–8
                b. CT commonly reveals intracranial lesion

**Table 1** Pediatric Glasgow Coma Scale (GCS)

| | 1 | 2 | 3 | 4 | 5 | 6 |
|---|---|---|---|---|---|---|
| Eyes | Does not open eyes | Opens eyes to pain | Opens eyes to speech | Opens eyes spontaneously | – | – |
| Verbal | No verbal response | Incomprehensible sounds | Inappropriate words | Confused | Oriented appropriate | – |
| Motor | No motor response | Extension to pain (decerebrate) | Abnormal flexion to pain (decorti-cate) | Withdraws from pain | Localizes painful stimulus | Obeys commands |

**Table 2** Infant Glasgow Coma Scale (GCS)

| | 1 | 2 | 3 | 4 | 5 | 6 |
|---|---|---|---|---|---|---|
| Eyes | Does not open eyes | Opens eyes to pain | Opens eyes to speech | Opens eyes spontaneously | – | – |
| Verbal | No verbal response | Moans to pain | Cries to pain | Irritable cries | Coos and babbles | – |
| Motor | No motor response | Extension to pain (decerebrate) | Flexion to pain (decorticate) | Withdraws from pain | Withdraws from touch | Moves spontaneously/purposefully |

**Table 3** Classification of traumatic brain injury (TBI)

| | GCS | Loss of consciousness |
|---|---|---|
| Mild | 13–15 | 0–30 min |
| Moderate | 9–12 | >30 min and <24 h |
| Severe | 3–8 | >24 h |

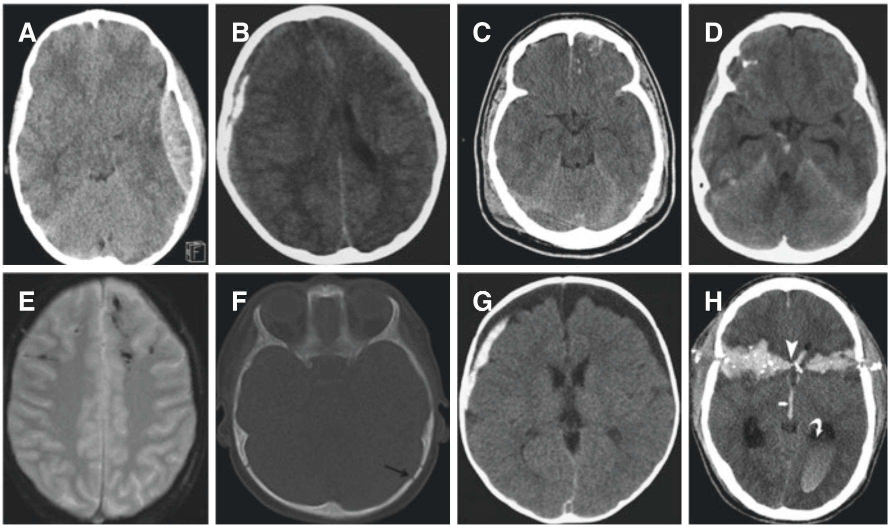

**Fig. 1** Types of traumatic brain injury. (A) Non-contrast computed tomography of the head showing left frontoparietal epidural hematoma (EDH) [2]. (B) Non-contrast computed tomography of the head showing right frontoparietal subdural hematoma with both acute and subacute components [3]. (C) Non-contrast computed tomography of the head showing left frontal contusion [4]. (D) Non-contrast computed tomography of the head showing intracranial hemorrhage in multiple compartments. Note traumatic subarachnoid hemorrhage within the basal cisterns [3]. (E) Magnetic resonance imaging (MRI), gradient echo sequence of the brain showing diffuse axonal injury seen in bilateral (left greater than right) frontal lobes [5]. (F) Non-contrast computed tomography of the head showing non-displaced skull fracture of left occipital bone just adjacent to lambdoid suture (arrow) [6]. (G) Non-contrast computed tomography of the head showing evidence of non-accidental trauma. Patients can present with subdural collections of various ages. Bilateral frontal hypodensities indicative of subdural hygromas. However, there is a hyperdensity on the right side indicative of acute subdural hematoma [3]. (H) Non-contrast computed tomography of the head showing penetrating head injury. Large arrowhead shows course of high velocity projectile across hemispheres. Small arrow reveals intraventricular hemorrhage within third ventricle. Curved arrow reveals intraventricular hemorrhage within the occipital horn of the left lateral ventricle [7]

c. Types of Traumatic Brain Injury (see Fig. 1)

  i. Epidural Hematoma (EDH) (see Fig. 1A)

    1. Hematoma contained within the epidural space.
    2. Can present with a lucid interval in 10–30% [2, 8] of patients, after which patients can rapidly deteriorate.
    3. Commonly associated with overlying skull fracture.

4. The origin of the hematoma is commonly arterial but can be venous in some cases.
5. Most commonly found in the temporal and parietal lobes.

ii. Subdural Hematoma (SDH) (see Fig. 1B)

  1. Hematoma found within the subdural space
  2. More commonly found in infants compared to toddlers and adolescents [9, 10]

iii. Intracerebral Hematoma (ICH)/Contusion (see Fig. 1C)

  1. Hematoma within the brain parenchyma
  2. Can be caused by coup/contrecoup injuries as a result of rapid acceleration and deceleration
  3. Commonly found in the frontal and temporal lobes due to proximity of skull base [11]

iv. Subarachnoid Hematoma (SAH) (see Fig. 1D)

  1. Hematoma within the subarachnoid space
  2. Can be found in 7.5% [12] of all pediatric TBI cases and up to 42% [13] of severe pediatric TBI cases
  3. Can be an independent prognostic indicator of outcomes, where SAH is associated with increased morbidity [13]

v. Diffuse Axonal Injury (DAI) (see Fig. 1E)

  1. Shearing injury of axons due to angular acceleration and deceleration.
  2. Most commonly found at the gray/white matter junction, corpus callosum, and brainstem.
  3. The Adams classification categorizes diffuse axonal injury based on pathology from histologic slides and clinical examination [14, 15].

    a. Grade 1 (mild) injury with microscopic changes in white matter within cerebral cortex, corpus callosum, and brainstem.
    b. Grade 2 (moderate) injury involves lesions within the corpus callosum in addition to the cerebral hemispheres.
    c. Grade 3 (severe) injury with grade 2 findings plus lesions in the brainstem, corpus callosum, and cerebral hemispheres.
    d. Studies have shown an unfavorable outcome in 17% of grade 1 patients, 40% for grade 2, and 62% for grade 3 [63, 64].

vi. Skull Fracture (see Fig. 1F)

  1. Can be classified as

    a. Closed (simple) vs open (compound)

- i. Closed skull fractures have no structural change to the overlying skin.
- ii. Open skull fractures have open defect of the overlying skin.
    1. Puts the patient at risk of infection if wound is grossly contaminated

b. Linear vs diastasis vs depressed

- i. Linear skull fractures are thin lines through the calvaria with no displacement of the natural bony contour.
- ii. Diastasis skull fractures are widening of the normal suture lines due to traumatic force.
- iii. Depressed skull fractures are breaks in calvarium in which one portion is displaced downward toward the underlying brain.
- iv. Growing skull fracture (posttraumatic leptomeningeal cyst) is a fracture with an underlying dural injury where the fracture appears to "grow" over time due to CSF pulsations eroding the bone.
- v. Ping-Pong fracture (green-stick fracture) is the caving of a focal area of the skull that resembles the crushed area of a ping-pong ball.
    1. Usually affects newborns due to plasticity of skull

c. Basilar

- i. Fractures of the skull base structures usually due to extremely high force
- ii. Can be associated with cerebrospinal fluid (CSF) leak (CSF otorrhea or rhinorrhea), vascular injury to the carotid and/or vertebral artery, cranial nerve injuries

d. Craniofacial

- i. Frontal sinus fracture
    1. Pneumatization of the frontal sinus starts at age 2 and continues throughout adolescence [16]
    2. Fracture patterns are based on the degree of involvement of the anterior and posterior tables.
    3. Fracture patterns in the pediatric population differ compared to adults due to children having smaller face-to-head volume ratio, thicker layer of adipose tissue, more malleable bones, and more flexible suture lines [16–19]
        - a. These differences require a greater amount of force to fracture the frontal sinus.

- vii. Non-accidental Head Trauma (see Fig. 1G)
    1. Indicative of child abuse.
    2. The CDC estimates that over 650,000 children were the victims of abuse or neglect in 2016.
        - a. Non-accidental trauma disproportionately affects infants [20].
            - i. Incidence in infants is 59 per 100,000 compared to 6 per 100,000 in children [20, 21].
    3. The following findings are suggestive of non-accidental trauma:
        - a. Multiple skull fractures
        - b. Associated intracranial injury
        - c. Chronic subdural hematomas (especially bilaterally)
        - d. Retinal hemorrhages
        - e. Multiple injuries of various ages in different locations
- viii. Penetrating Head Injury (see Fig. 1H)
    1. Usually caused by high-velocity projectiles such as bullets from gunshot. Can be caused by low-velocity objects such as knives.
    2. Injury cascade is classified as
        - a. Primary Injury
            - i. Due to direct trauma to the skull, scalp, and surrounding soft tissue from the projectile object.
            - ii. The extent of injury is related to the mass and velocity of the object.
        - b. Secondary Injury
            - i. Due to subsequent edema and increased ICP.
            - ii. Brain hemodynamics can adversely affect cardiac function and coagulation (DIC).

- d. Primary Injury
    - i. Due to direct glial disruption, axon shearing, and vascular injuries
- e. Secondary Injury
    - i. Due to hypoxia, hypotension, hypovolemia, systemic trauma, electrolyte abnormalities, hyperthermia, and infection.
    - ii. Goal is to minimize secondary injury by optimizing cerebral perfusion pressure (CPP) and oxygenation.

1. CPP = Mean Arterial Pressure (MAP) – Intracranial Pressure (ICP)
    a. Normal CPP in adult patients ranges between 50 and 70. In pediatric patients, normal CPP is not well established but assumed to be less due to lower MAP and ICP.

2. Monitoring

a. Intracranial Pressure (ICP)
    i. The use of ICP monitoring in pediatric patients with severe TBI is suggested (Level III evidence [22]).
        1. Cerebral edema occurs within the closed compartment of the skull as a result of secondary injury to the brain. This leads to increased ICP which places patients at risk of herniation syndromes, further cerebral edema, ischemic changes, and death [22]. Thus, ICP should be closely monitored as changes can dictate treatment algorithms (see below).
        2. Evidence shows that pediatric patients with severe TBI are at notable risk of developing intracranial hypertensions [23, 24]. There are no particular markers that indicate the presence or absence of intracranial hypertension, thus making monitoring critical.
        3. ICP data is useful as a prognostic variable in the management of pediatric patients with severe TBI [22, 25, 26].
        4. ICP monitoring and treatment have been found to be associated with a reduction in mortality associated with intracranial hypertension [27–29].
    ii. ICP monitoring is NOT routinely used in pediatric patients with mild or moderate TBI.

b. Advanced Neuromonitoring
    i. Use of advanced neuromonitoring (e.g., brain oxygenation) should only be used in patients with no contraindications to invasive neuromonitoring (i.e., coagulopathy) and without diagnosis of brain death (Level III evidence).
    ii. If brain tissue oxygenation is used, it is suggested to maintain a level greater than 10 mmHg (Level III evidence [22]).
        1. Advanced neuromonitoring provides information about the cerebrovascular and metabolic functions in the brain and possible treatments.
        2. Evidence reveals that brain oxygenation greater than 25 mmHg is associated with more favorable outcomes [30] while oxygenation less than 10 mmHg is associated with worse outcomes [31].

c. Neuroimaging

    i. It is suggested to not exclude the possibility of elevated ICP in comatose pediatric patients on the basis of a normal initial (0–6 h post-injury) CT examination (Level III evidence [22])

        1. Evidence showed elevated ICP >20 in a majority of a small population of pediatric patients with initial normal head CT [32].

    ii. Routine CT scans greater than 24 h after admission and initial follow-up are not suggested for decision-making in regard to neurosurgical intervention, unless there is evidence of neurologic deterioration or increasing ICP (Level III evidence [22]).

        1. Multiple studies show that repeat imaging did not affect the decision making for any neurosurgical intervention in patient with no neurologic change[33, 34]

    iii. CT imaging of the head allows for rapid detection of intracranial injury, mass lesions, and signs of intracranial hypertension.

        1. The severity of the intracranial injury seen on CT imaging can be measured using either the Rotterdam CT score [35] or Marshall classification system [36]. Both assess the degree of swelling (due to secondary injury) by assessing midline shift and/or compression of the basal cisterns. The two differ as the Marshall system looks at “high or mixed density lesions,” where the Rotterdam looks for the presence of epidural, subarachnoid, or intraventricular hemorrhage.

3. Thresholds

    a. Intracranial Pressure (ICP)

        i. Treatment of elevated ICP should target threshold <20 mmHg (Level III evidence [22]).
        ii. The majority of mortality from severe TBI is due to refractory intracranial hypertension with evidence showing increased mortality with ICP >20 mmHg.
        iii. As the ICP rises, CPP decreases, leading to further secondary injury.
        iv. Normal ICP values are lower in the pediatric population compared to adults. Even though guidelines recommend a target threshold of 20, an individualized treatment plan with lower thresholds may be physiologically appropriate for younger children and infants.

            1. There is debate on which threshold should be used across different age groups within the pediatric population.

                a. There have been described thresholds of 15 mmHg for age 0–24 months; 18 mmHg for age 25–96 months; and 20 mmHg for age 97–156 months [37].

v. Stepwise progression for treatment of elevated ICP.
    a. Raise HOB >30.
        i. Reduces ICP by improving venous outflow
    b. Keep neck straight; avoid neck constriction.
        i. Kinking of jugular vein outflow increases ICP.
    c. Avoid arterial hypotension (SBP <90 mmHg).
        i. Hypotension reduces cerebral blood flow.
    d. Control hypertension.
    e. Avoid hypoxia.
        i. Can further worsen ischemic injury
    f. Ventilate for normocarbia.
    g. Pharmacologic sedation.
        i. Reduces sympathetic tone, elevated blood pressure from movement, pain
    h. Drain CSF if intraventricular catheter is present.
        i. Decreases intracranial volume (due to Kellie-Monroe doctrine)
    i. Barbiturate therapy.
        i. Decreases metabolic demand and cerebral blood flow leading to decreased ICP
    j. Hyperosmolar therapy.
        i. Increased osmolarity decreases cerebral swelling, decrease
    k. Hyperventilation.
        i. Decreased $CO_2$ leads to vasoconstrictions of the vasculature, leading to decreased blood volume and decreased ICP.
            1. Usually reserved as a temporary last resort (bridging therapy) until more definitive treatments can be undertaken
    l. Decompressive craniectomy
        i. Removes rigid fixed skull

b. Cerebral Perfusion Pressure (CPP)
    i. CPP=mean arterial pressure (MAP) – intracranial pressure (ICP). Accurate measurement of ICP requires invasive monitoring.
    ii. A target CPP of 40–50 mm Hg is suggested, although consideration should be made age-specific accounting for lower threshold in infants and higher thresholds in adolescents (Level III evidence [22])

            1. Evidence shows that patients with CPP less than 40 mmHg were more likely to have unfavorable outcomes while patients with CPP greater than 40 mmHg [31, 38] were found to have more favorable outcomes [25, 38].

        iii. CPP provides the pressure gradient for cerebral blood flow (CBF) into the brain. Under normal circumstances, CBF is under the control of autoregulation. In autoregulation, CBF is maintained despite changes in cerebral vascular resistance and CPP.

            1. Autoregulation can be disrupted in TBI, where a decrease in CPP can lead to marked changes in CBF and thus ischemic changes.
            2. Close monitoring of ICP, MAP, and CPP allows an opportunity to prevent neurologic injury.

4. Treatments

    a. Hyperosmolar Therapy

        i. Hypertonic saline

            1. Hypertonic saline reduces ICP by creating an osmotic gradient as sodium cannot easily cross the blood-brain barrier.
            2. Bolus hypertonic saline (3%) is recommended in patients with intracranial hypertension with recommended dose between 2 and 5 mL/kg over 10–20 min (Level II evidence [22]).

                a. Multiple studies show the quick and reliable effect of hypertonic saline on reducing ICP in pediatric patients with severe TBI [39, 40].

            3. Continuous hypertonic saline is suggested in patients with intracranial hypertension with recommended dose between 0.1 and 1 mL/kg per hour (Level III evidence [22]).

                a. Randomized controlled trials have compared hypertonic saline 1.7% vs lactated Ringer's and showed pediatric patients with hypertonic saline required fewer interventions to control ICP. The study was limited due to its use of both lactated Ringer's and 1.7% hypertonic saline, which has limited clinical use in pediatrics [41].

            4. Bolus 23.4% hypertonic saline is suggested for refractory ICP with recommended dose of 0.5 mL/kg and maximum dose of 30 mL (Level III evidence [22]).

                a. Evidence has shown that 23.4% hypertonic saline can be used in cases of refractory intracranial hypertension and actually led to a mean reduction in ICP of 10 mmHg and more favorable outcome [42].

    ii. Mannitol
        1. Mannitol has been used to reduce ICP in children for decades but has not been studied in any recent controlled clinical trial in a pediatric population only. Regardless, mannitol is widely used for management of refractory ICP in intracranial hypertension.
        2. Mannitol reduces ICP via two mechanisms:
            a. Reduces blood viscosity
            b. Osmotic effect

b. Analgesic, Sedatives, Neuromuscular Blockade
    i. If a patient has adequate and appropriate use of analgesia and sedation with multiple ICP-related therapies, it's suggested to avoid bolus administration of fentanyl and/or midazolam during ICP crises due to risk of cerebral hypoperfusion (Level III evidence).
    ii. Analgesics
        1. Fentanyl is a highly lipid-soluble narcotic.
        2. Used in combination with midazolam for sedation and to reduce ICP when related to pain and discomfort.
    iii. Sedatives
        1. Midazolam is a benzodiazepine used in multiple scenarios.
        2. Used in combination with fentanyl for sedation and to reduce ICP related to pain and discomfort.
    iv. Neuromuscular blockade
        1. Reduces ICP by optimizing the patient-ventilator interactions and decreasing shivering
    v. Ketamine
        1. Traditionally, ketamine has been contraindicated in patients with increased ICP due to concerns of ketamine increasing CBF. Studies are ongoing on the validity of its effects on cerebral dynamics [43].

c. CSF Drainage
    i. CSF drainage through an external ventricular drain (EVD) can be used to manage increased ICP (Level III evidence).
        1. CSF drainage decreases the total cerebral volume, per the Kellie-Monroe doctrine ($\mathrm{Vol}_{\mathrm{Brain}} + \mathrm{Vol}_{\mathrm{Blood}} + \mathrm{Vol}_{\mathrm{CSF}}$).

d. Seizure Prophylaxis
    i. Seizure prophylaxis is recommended in TBI patient to reduce incidence of early posttraumatic seizures (Level III evidence).

- ii. Posttraumatic seizures can be classified as
    1. Early: occur 7 days after injury
    2. Late: occur after 7 days from the injury
- iii. Risk factors for posttraumatic seizures include
    1. Location of lesion
    2. Intraparenchymal contusion
    3. Retained foreign objects, bone
    4. Depressed skull fractures
    5. Penetrating injury
- iv. Can use prophylaxis with either
    1. Phenytoin
    2. Levetiracetam

e. Ventilation Therapies

- i. Hyperventilation reduces ICP by decreasing the amount of $CO_2$, leading to cerebral vasoconstriction and subsequent reduction in CBF and CBV.
- ii. Prophylactic severe hyperventilation to a $PaCO_2$ less than 30 mmHg in the initial 48 h after injury is not recommended (Level III evidence).
    1. Subsequent studies showed that the decrease in CBF from hyperventilation was associated with unfavorable outcomes and can lead to ischemia [44, 45].
- iii. If hyperventilation is used in management of refractory intracranial hypertension, advanced neuromonitoring for monitoring of cerebral ischemia is recommended (Level III evidence [22]).

f. Temperature Control

- i. Posttraumatic hypothermia is defined as core body temperature less than 35 °C.
- ii. Hyperthermia is defined as core body temperature greater than 38.0–38.5 °C.
    1. Hyperthermia can worsen secondary brain injury as it increases cerebral metabolic demand, inflammation, excitotoxicity, and cell death.
- iii. Prophylactic moderate (32–33 °C) hypothermia is not recommended over normothermia to improve outcomes (Level II evidence [22]).
    1. Multiple studies have shown that there was no change in mortality or outcomes in patient who underwent hypothermia vs normothermia therapies [46–48].

- iv. Moderate (32–33 °C) hypothermia is suggested for ICP control (Level III evidence [22]).
    1. Studies show significantly lower ICP in pediatric patients who underwent cooling compared to controls but only as an immediate effect [37, 49].
    2. Care must be taken during rewarming, as it should be carried out at rate of 0.5–1.0 °C every 12–24 h to avoid complication.
    3. Care must be taken during phenytoin use as dosing should be adjusted to minimize toxicity, specifically during rewarming process.

- g. Barbiturate Therapy
  - i. Barbiturates lower ICP by decoupling metabolic demands from CBF, which results in improved oxygenation, decreased CBF, and decreased CBV.
    1. Pentobarbital is the most common used barbiturate in children.
    2. Dosing is maintained to achieve burst suppression, and thus continuous electroencephalogram (EEG) is required to monitor optimal dosing.
    3. Cardiorespiratory complications are common when using barbiturates.
  - ii. High-dose barbiturate therapy is suggested in hemodynamically stable patients with refractory intracranial hypertension despite maximal medical and surgical therapy (Level III evidence [22])
- h. Decompressive Craniectomy
  - i. Decompressive craniectomy can be classified as
    1. Incidental (Primary): performed concomitantly with removal of mass lesion
    2. Therapeutic (Secondary): performed solely for refractory intracranial hypertension
       - a. Refractory intracranial hypertension can be classified as early (within 72 h of admission) or late (within 10 days of admission).
  - ii. Decompressive craniectomy is recommended to treat neurologic deterioration, herniation, or intracranial hypertension refractory to medical management (Level III evidence [22]).
    1. Multiple studies show that decompressive craniectomy is effective at reducing ICP in patients refractory to medical management [50–52].
  - iii. There is conflicting evidence on the effect of decompressive craniectomy on mortality and outcome.
    1. Controversy is due to lack of clearly defined indications, its timing, lack of optimal surgical technique, differences in outcomes, and significant risk of complications [22]

        a. In the adult population, the results of the RESCUEicp [53] and DECRA [54] trials have addressed some concerns and led to the development of Level IIa recommendations [55]:
            i. Secondary decompressive craniectomy for late refractory ICP elevation is recommended to improve mortality and favorable outcomes.
            ii. Secondary decompressive craniectomy for early refractory ICP elevation is NOT recommended to improve mortality and favorable outcomes.
            iii. A large frontotemporoparietal decompressive craniectomy (at least 12 × 15 cm or 15 cm in diameter) is recommended over a small frontotemporoparietal decompressive craniectomy to reduce mortality and improve neurological outcome.
            iv. Secondary decompressive craniectomy, performed for either early or late refractory ICP elevation is suggested to reduce the length of stay in the ICU, although the relationship between these effects on outcomes is unclear.
        b. In the pediatric population, some studies state no difference was seen between patients who underwent surgery and those who had maximum medical management [50, 56, 57]. Other studies have shown a decrease in mortality and improved outcomes after decompressive craniectomy [58, 59].

i. Nutrition
    i. TBI results in an increase in system-wide metabolism with subsequent need for increased caloric support.
    ii. Hyperglycemia occurs in response to the constant stress from the initial injury.
        1. The influence of strict glucose control remains controversial in both the adult and pediatric population. Due to lack of consistency of findings, there are no formal recommendations about glucose control in both adult [60] and pediatric [22] populations.
    iii. Use of an immune-modulating diet is not recommended (Level II evidence [22]).
        1. No difference in outcome was noted in pediatric patients who received immune-enhancing diet versus regular formula [61].
    iv. Early enteral nutritional support (within 72 h from injury) is suggested to be initiated to decrease mortality and improve outcomes (Level III evidence [22]).
        1. Pediatric patients with TBI commonly have polytrauma that necessitates parenteral nutrition.

j. Steroids
    i. Corticosteroids work by restoring vascular permeability, inhibiting tumor-induced angiogenesis, and decreasing edema and CSF production.
        1. Despite mechanism of action, studies have shown steroid use to be associated with worsened outcomes.
    ii. The use of corticosteroids is not suggested to improve outcome or reduce ICP (Level III evidence [22]).
        1. Studies show that steroids do not have any effect on ICP but also increase risk of other complications due to its use in pediatric population [62].

## Study Questions

1. All of the following are suggestive of non-accidental trauma EXCEPT
    a. Multiple skull fractures
    b. Bilateral chronic subdural hematomas
    c. Normal retinal exam
    d. Multiple injuries of various ages in different locations

    Answer: C. Normal retinal exam. Findings suggestive of non-accidental trauma include multiple skull fractures, bilateral chronic subdural hematomas, multiple injuries of various ages, and retinal hemorrhages.

2. Which of the following is NOT a risk factor for posttraumatic seizure?
    a. Retained foreign object, bone
    b. Linear skull fracture
    c. Intraparenchymal contusion
    d. Penetrating injury

    Answer: B. Linear skull fracture. A linear skull fracture is NOT a risk factor for posttraumatic seizure. However, a depressed skull fracture is a risk factor for posttraumatic seizure due to depression and mass effect from the depressed bone.

## References

1. Carlson K, Kehle S, Meis L, et al. VA Evidence-based synthesis program reports. In: The assessment and treatment of individuals with history of traumatic brain injury and post-traumatic stress disorder: a systematic review of the evidence. Washington, DC: Department of Veterans Affairs (US); 2009. https://www.ncbi.nlm.nih.gov/books/NBK49144/. Accessed 4 Apr 2021.

2. Spazzapan P, Krašovec K, Velnar T. Risk factors for bad outcome in pediatric epidural hematomas: a systemic review. Chinese Neurosurg J. 2019;5(1):1–9.
3. Boychuk RB. Radiology. In Ped Emerg Med, Vol 4, Case 2. Radiol Cases Pediatr Emerg Med. 4. https://www.hawaii.edu/medicine/pediatrics/pemxray/v5c07.html. Accessed 25 May 2021.
4. Cerebral Contusion | Pediatric Radiology Reference Article | PediatricImaging.org | @pedsimaging. https://pediatricimaging.org/diseases/cerebral-contusion/. Accessed 25 May 2021.
5. Parizel PM, Van Goethem JW, Özsarlak Ö, Maes M, Phillips CD. New developments in the neuroradiological diagnosis of craniocerebral trauma. Eur Radiol. 2005;15(3):569–81.
6. Orman G, Wagner MW, Seeburg D, et al. Pediatric skull fracture diagnosis: should 3D CT reconstructions be added as routine imaging? J Neurosurg Pediatr. 2015;16(4):426–31.
7. Bodanapally UK, Saksobhavivat N, Shanmuganathan K, Aarabi B, Roy AK. Arterial injuries after penetrating brain injury in civilians: risk factors on admission head computed tomography. J Neurosurg. 2015;122(1):219–26.
8. Paşaoglu A, Orhon C, Koç K, Selçuklu A, Akdemir H, Uzunoglu H. Traumatic extradural haematomas in pediatric age group. Acta Neurochir. 1990;106(3-4):136–9.
9. Proctor M. Intracranial subdural hematoma in children: clinical features, evaluation, and management. UpToDate; 2014. https://www.uptodate.com/contents/intracranial-subdural-hematoma-in-children-clinical-features-evaluation-and-management#references. Accessed 25 May 2021.
10. Datta S, Stoodley N, Jayawant S, Renowden S, Kemp A. Neuroradiological aspects of subdural haemorrhages. Arch Dis Child. 2005;90(9):947–51.
11. Faried A, Halim D, Widjaya IA, Badri RF, Sulaiman SF, Arifin MZ. Correlation between the skull base fracture and the incidence of intracranial hemorrhage in patients with traumatic brain injury. Chin J Traumatol. 2019;22(5):286–9.
12. Dalle Ore CL, Rennert RC, Schupper AJ, et al. The identification of a subgroup of children with traumatic subarachnoid hemorrhage at low risk of neuroworsening. J Neurosurg Pediatr. 2018;22(5):559–66.
13. Hochstadter E, Stewart TC, Alharfi IM, Ranger A, Fraser DD. Subarachnoid hemorrhage prevalence and its association with short-term outcome in pediatric severe traumatic brain injury. Neurocrit Care. 2014;21(3):505–13.
14. Adams JH, Doyle D, Ford I, Gennarelli TA, Graham DI, Mclellan DR. Diffuse axonal injury in head injury: definition, diagnosis and grading. Histopathology. 1989;15(1):49–59.
15. Maxwell WL, Graham DI. Diffuse axonal injury (DAI). In: Encyclopedia of the neurological sciences. Elsevier; 2003. p. 10–2.
16. Vu AT, Patel PA, Chen W, Wilkening MW, Gordon CB. Pediatric frontal sinus fractures: outcomes and treatment algorithm. J Craniofac Surg. 2015;26(3):776–81.
17. Alcalá-Galiano A, Arribas-García IJ, Martin-Pérez MA, Romance A, Montalvo-Moreno JJ, Juncos JMM. Pediatric facial fractures: children are not just small adults. Radiographics. 2008;28(2):441–61.
18. Cole P, Kaufman Y, Hollier LH. Managing the pediatric facial fracture. Craniomaxillofac Trauma Reconstr. 2009;2(2):77–83.
19. Ryan ML, Thorson CM, Otero CA, et al. Pediatric facial trauma: a review of guidelines for assessment, evaluation, and management in the emergency department. J Craniofac Surg. 2011;22(4):1183–9.
20. Delaplain PT, Grigorian A, Won E, et al. Nonaccidental trauma is an independent risk factor for mortality among injured infants. Pediatr Emerg Care. 2019.
21. Leventhal JM, Martin KD, Gaither JR. Using US data to estimate the incidence of serious physical abuse in children. Pediatrics. 2012;129(3):458–64. https://doi.org/10.1542/peds.2011-1277.
22. Kochanek PM, Tasker RC, Carney N, et al. Guidelines for the management of pediatric severe traumatic brain injury, third edition: update of the brain trauma foundation guidelines. Pediatr Crit Care Med. 2019;20(3):S1–S82.

23. Barzilay Z, Augarten A, Sagy M, Shahar E, Yahav Y, Boichis H. Variables affecting outcome from severe brain injury in children. Intensive Care Med. 1988;14(4):417–21.
24. Shapiro K, Marmarou A. Clinical applications of the pressure-volume index in treatment of pediatric head injuries. J Neurosurg. 1982;56(6):819–25.
25. Chambers IR, Treadwell L, Mendelow AD. Determination of threshold levels of cerebral perfusion pressure and intracranial pressure in severe head injury by using receiver-operating characteristic curves: an observational study in 291 patients. J Neurosurg. 2001;94(3):412–6.
26. Alberico AM, Ward JD, Choi SC, Marmarou A, Young HF. Outcome after severe head injury. Relationship to mass lesions, diffuse injury, and ICP course in pediatric and adult patients. J Neurosurg. 1987;67(5):648–56.
27. Alkhoury F, Kyriakides TC. Intracranial pressure monitoring in children with severe traumatic brain injury: national trauma data bank-based review of outcomes. JAMA Surg. 2014;149(6):544–8.
28. Bennett TD, Riva-Cambrin J, Keenan HT, Korgenski EK, Bratton SL. Variation in intracranial pressure monitoring and outcomes in pediatric traumatic brain injury. Arch Pediatr Adolesc Med. 2012;166(7):641–7.
29. Bennett TD, DeWitt PE, Greene TH, et al. Functional outcome after intracranial pressure monitoring for children with severe traumatic brain injury. JAMA Pediatr. 2017;171(10):965–71.
30. Stippler M, Ortiz V, Adelson PD, et al. Brain tissue oxygen monitoring after severe traumatic brain injury in children: relationship to outcome and association with other clinical parameters. J Neurosurg Pediatr. 2012;10(5):383–91.
31. Figaji AA, Zwane E, Thompson C, et al. Brain tissue oxygen tension monitoring in pediatric severe traumatic brain injury: Part 1: relationship with outcome. Childs Nerv Syst. 2009;25(10):1325–33.
32. Bailey BM, Liesemer K, Statler KD, Riva-Cambrin J, Bratton SL. Monitoring and prediction of intracranial hypertension in pediatric traumatic brain injury: clinical factors and initial head computed tomography. J Trauma Acute Care Surg. 2012;72:263–70.
33. Bata SC, Yung M. Role of routine repeat head imaging in paediatric traumatic brain injury. ANZ J Surg. 2014;84(6):438–41.
34. Figg RE, Stouffer CW, Vander Kolk WE, Connors RH. Clinical efficacy of serial computed tomographic scanning in pediatric severe traumatic brain injury. Pediatr Surg Int. 2006;22(3):215–8.
35. Liesemer K, Riva-Cambrin J, Bennett KS, et al. Use of rotterdam CT scores for mortality risk stratification in children with traumatic brain injury. Pediatr Crit Care Med. 2014;15(6):554–62.
36. Marshall LF, Marshall SB, Klauber MR, et al. The diagnosis of head injury requires a classification based on computed axial tomography. J Neurotrauma. 1992;9(SUPPL. 1):S287–92. https://europepmc.org/article/med/1588618. Accessed 5 Apr 2021
37. Adelson PD, Ragheb J, Muizelaar JP, et al. Phase II clinical trial of moderate hypothermia after severe traumatic brain injury in children. Neurosurgery. 2005;56(4):740–53.
38. Downard C, Hulka F, Mullins RJ, et al. Relationship of cerebral perfusion pressure and survival in pediatric brain-injured patients. J Trauma: Inj Inf Crit Care. 2000;49:654–9.
39. Shein SL, Ferguson NM, Kochanek PM, et al. Effectiveness of pharmacological therapies for intracranial hypertension in children with severe traumatic brain injury-results from an automated data collection system time-synched to drug administration. Pediatr Crit Care Med. 2016;17(3):236–45.
40. Peterson B. Hypertonic saline lowers raised intracranial pressure in children after head trauma. J Neurosurg Anesthesiol. 1992;4(1):4–10.
41. Simma B, Burger R, Falk M, Sacher P, Fanconi S. A prospective, randomized, and controlled study of fluid management in children with severe head injury: Lactated ringer's solution versus hypertonic saline. Crit Care Med. 1998;26(7):1265–70.
42. Piper BJ, Harrigan PW. Hypertonic saline in paediatric traumatic brain injury: a review of nine years' experience with 23.4% hypertonic saline as standard hyperosmolar therapy. Anaesth Intensive Care. 2015;43(2):204–10.

43. Bar-Joseph G, Guilburd Y, Tamir A, Guilburd JN. Effectiveness of ketamine in decreasing intracranial pressure in children with intracranial hypertension: clinical article. J Neurosurg Pediatr. 2009;4(1):40–6.
44. David Adelson P, Srinivas R, Chang Y, Bell M, Kochanek PM. Cerebrovascular response in children following severe traumatic brain injury. Childs Nerv Syst. 2011;27(9):1465–76.
45. Adelson PD, Clyde B, Kochanek PM, Wisniewski SR, Marion DW, Yonas H. Cerebrovascular response in infants and young children following severe traumatic brain injury: a preliminary report. Pediatr Neurosurg. 1997;26(4):200–7. https://doi.org/10.1159/000121192.
46. Crompton EM, Lubomirova I, Cotlarciuc I, Han TS, Sharma SD, Sharma P. Meta-analysis of therapeutic hypothermia for traumatic brain injury in adult and pediatric patients*. Crit Care Med. 2017;45(4):575–83.
47. Tasker RC, Akhondi-Asl A. Updating evidence for using therapeutic hypothermia in pediatric severe traumatic brain injury. Crit Care Med. 2017;45(10):e1091.
48. Tasker RC, Vonberg FW, Ulano ED, Akhondi-Asl A. Updating evidence for using hypothermia in pediatric severe traumatic brain injury: conventional and Bayesian meta-analytic perspectives. Pediatr Crit Care Med. 2017;18(4):355–62. https://doi.org/10.1097/PCC.0000000000001098.
49. Hutchison JS, Ward RE, Lacroix J, et al. Hypothermia therapy after traumatic brain injury in children. N Engl J Med. 2008;358(23):2447–56.
50. Taylor A, Butt W, Rosenfeld J, et al. A randomized trial of very early decompressive craniectomy in children with traumatic brain injury and sustained intracranial hypertension. Childs Nerv Syst. 2001;17(3):154–62.
51. Cho DY, Wang YC, Chih CS. Decompressive craniotomy for acute shaken/impact baby syndrome. Pediatr Neurosurg. 1995;23(4):192–8.
52. Figaji AA, Fieggen AG, Peter JC. Early decompressive craniotomy in children with severe traumatic brain injury. Childs Nerv Syst. 2003;19(9):666–73.
53. Hutchinson PJ, Kolias AG, Timofeev IS, et al. Trial of decompressive craniectomy for traumatic intracranial hypertension. N Engl J Med. 2016;375(12):1119–30.
54. Cooper DJ, Rosenfeld JV, Murray L, et al. Patient outcomes at twelve months after early decompressive craniectomy for diffuse traumatic brain injury in the randomized DECRA clinical trial. J Neurotrauma. 2020;37(5):810–6.
55. Hawryluk GWJ, Rubiano AM, Totten AM, et al. Guidelines for the management of severe traumatic brain injury: 2020 update of the decompressive craniectomy recommendations. Neurosurgery. 2020;87(3):427–34.
56. Thomale UW, Graetz D, Vajkoczy P, Sarrafzadeh AS. Severe traumatic brain injury in children-a single center experience regarding therapy and long-term outcome. Childs Nerv Syst. 2010;26(11):1563–73.
57. Mhanna MJ, El Mallah W, Verrees M, Shah R, Super DM. Outcome of children with severe traumatic brain injury who are treated with decompressive craniectomy. J Neurosurg Pediatr. 2015;16(5):508–14.
58. Josan VA, Sgouros S. Early decompressive craniectomy may be effective in the treatment of refractory intracranial hypertension after traumatic brain injury. Childs Nerv Syst. 2006;22(10):1268–74.
59. Rubiano AM, Villarreal W, Hakim EJ, et al. Early decompressive craniectomy for neurotrauma: an institutional experience. Ulus Travma ve Acil Cerrahi Derg. 2009;15(1):28–38.
60. Carney N, Totten AM, O'Reilly C, et al. Guidelines for the management of severe traumatic brain injury, Fourth Edition. Neurosurg. 2017;80(1):6–15.
61. Briassoulis G, Filippou O, Kanariou M, Papassotiriou I, Hatzis T. Temporal nutritional and inflammatory changes in children with severe head injury fed a regular or an immune-enhancing diet: a randomized, controlled trial. Pediatr Crit Care Med. 2006;7(1):56–62.
62. Fanconi S, Klöti J, Meuli M, Zaugg H, Zachmann M. Dexamethasone therapy and endogenous cortisol production in severe pediatric head injury. Intensive Care Med. 1988;14(2):163–6.

63. van Eijck MM, Herklots MW, Peluso J, Schoonman GG, Oldenbeuving AW, de Vries J, van der Naalt J, Roks G. Accuracy in prediction of long-term functional outcome in patients with traumatic axonal injury: a comparison of MRI scales. Brain Inj. 2020;34(5):595–601.
64. Gentry LR. Imaging of closed head injury. Radiology. 1994;191(1):1–17.

## *Further Reading*

Kochanek PM, Tasker RC, Carney N, et al. Guidelines for the management of pediatric severe traumatic brain injury, third edition: update of the brain trauma foundation guidelines. Pediatr Crit Care Med. 2019;20(3):S1–S82.

# Pediatric Head Trauma and Treatment Algorithm

**Cameron Brimley, George Jallo, and Nir Shimony**

**Key Points**

1. Traumatic brain injury is the leading cause of morbidity and mortality among pediatric patients.
2. Initial assessment and management with ATLS/PALS protocols are followed by performing neurologic assessments and obtaining immediate CT images to identify pathology that needs to be handled emergently.
3. In addition to the immediate surgical treatment for certain pathologies, management focuses on preventing elevated ICP while maintaining appropriate cerebral blood flow and oxygenation to prevent secondary brain injury.

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1. Background

    a. Head trauma is a broad category and includes a complex set of pathologies. Although extensive research has been completed in the area of pediatric head trauma including multiple attempts at multicenter randomized controlled trials, minimal level 1 evidence exists in guiding management. Guidelines and algorithms were published in 2019 as a third edition update to the Brain Trauma Foundation Guidelines compiling the most up-to-date available research with predominately level 2 and level 3 recommendations [1–3]. This chapter provides a concise summary of these guidelines and other recent literature providing a framework and algorithm for treatment of pediatric patients with head trauma.
    b. A significant portion of TBI management centers around ensuring adequate cerebral blood flow and oxygenation by maintaining cerebral perfusion pressure and normal intracranial pressure. First-line therapies include hypertonic/hyperosmolar therapy, decreasing $PaCO_2$, CSF drainage, increasing sedation, and neuromuscular blockade. Second-tier therapies include decompressive hemicraniectomy, barbiturate coma, and increased sedation. A pathway for management is detailed below in Fig. 1 [1, 2, 4].

2. Epidemiology

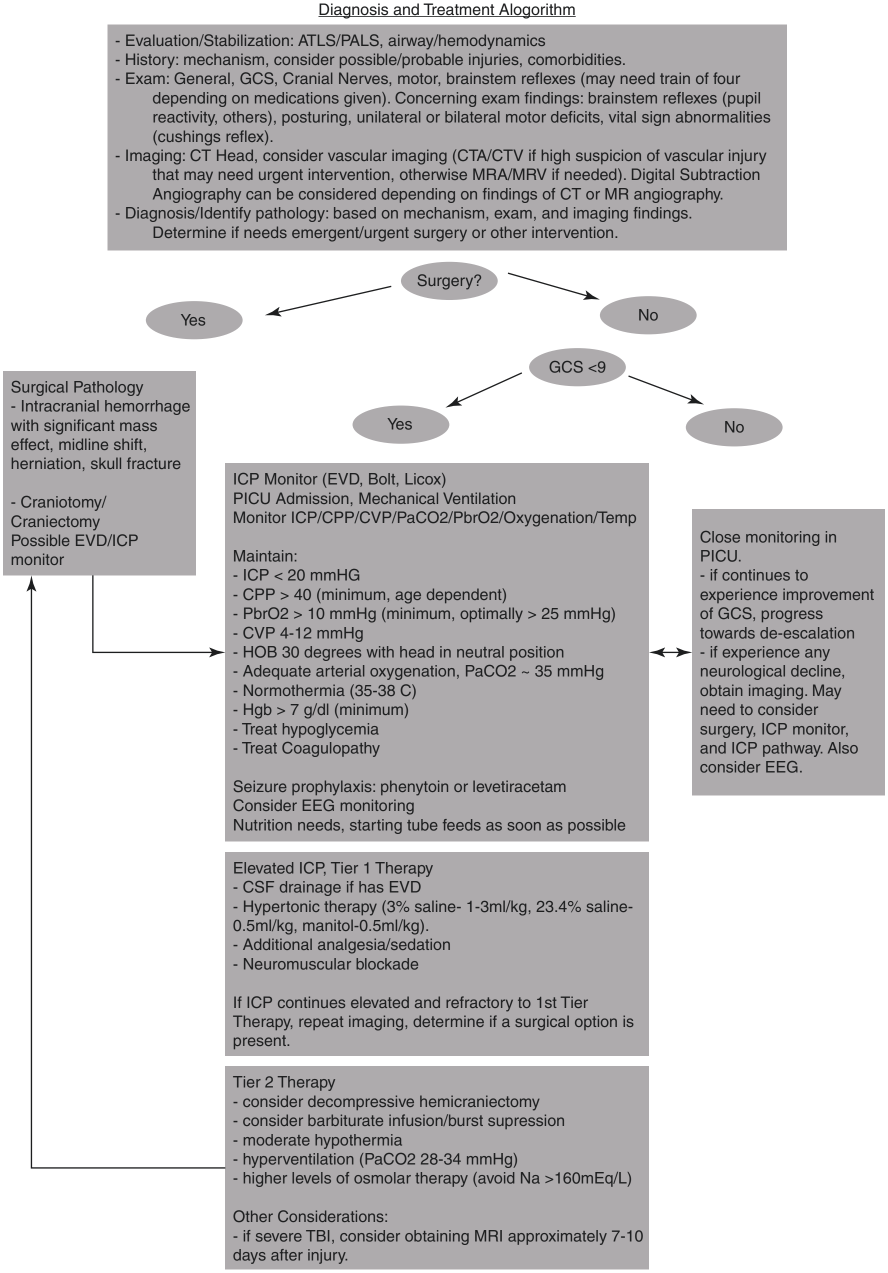
    a. Traumatic brain injury is the leading cause of morbidity and mortality among pediatric patients, with an estimated 280/100,000 annual incidence.
    b. Nearly 500,000 TBI events occur each year and account for more than 70% of fatal injuries to children.
    c. Children hospitalized with TBI have reported mortality upward of 24.2%.
    d. Survivors are subject to traumatic epilepsy, cognitive deficits, sleep disorders, and psychiatric disorders.
    e. Total life costs of injuries for children (<14 years old) with TBI estimated are at $60.4 billion [5, 6].

3. Pathophysiology

    a. Primary brain injury results from the initial traumatic event and includes the original damage to the brain/nerves, blood vessels, dura, bone, and soft tissues.
    b. Secondary brain injury can occur in the days to weeks after the event resulting from elevated intracranial pressure causing decreased cerebral blood flow with subsequent ischemic damage as well as causing cerebral herniation affecting the cranial nerves and brainstem [7–9].

4. Diagnosis

    a. Initial assessment and management with ATLS/PALS protocols are followed by performing neurologic assessments and obtaining immediate CT images to identify pathology that needs to be handled emergently. See Fig. 1.
    b. The neurological exam includes assessment of Glasgow Coma Scale (GCS) and assessment of cranial nerves, followed by motor and sensory examination.



Diagnosis and Treatment Alogorithm

- Evaluation/Stabilization: ATLS/PALS, airway/hemodynamics
- History: mechanism, consider possible/probable injuries, comorbidities.
- Exam: General, GCS, Cranial Nerves, motor, brainstem reflexes (may need train of four depending on medications given). Concerning exam findings: brainstem reflexes (pupil reactivity, others), posturing, unilateral or bilateral motor deficits, vital sign abnormalities (cushings reflex).
- Imaging: CT Head, consider vascular imaging (CTA/CTV if high suspicion of vascular injury that may need urgent intervention, otherwise MRA/MRV if needed). Digital Subtraction Angiography can be considered depending on findings of CT or MR angiography.
- Diagnosis/Identify pathology: based on mechanism, exam, and imaging findings. Determine if needs emergent/urgent surgery or other intervention.

Surgical Pathology
- Intracranial hemorrhage with significant mass effect, midline shift, herniation, skull fracture

- Craniotomy/ Craniectomy
Possible EVD/ICP monitor

ICP Monitor (EVD, Bolt, Licox)
PICU Admission, Mechanical Ventilation
Monitor ICP/CPP/CVP/PaCO2/PbrO2/Oxygenation/Temp

Maintain:
- ICP < 20 mmHG
- CPP > 40 (minimum, age dependent)
- PbrO2 > 10 mmHg (minimum, optimally > 25 mmHg)
- CVP 4-12 mmHg
- HOB 30 degrees with head in neutral position
- Adequate arterial oxygenation, PaCO2 ~ 35 mmHg
- Normothermia (35-38 C)
- Hgb > 7 g/dl (minimum)
- Treat hypoglycemia
- Treat Coagulopathy

Seizure prophylaxis: phenytoin or levetiracetam
Consider EEG monitoring
Nutrition needs, starting tube feeds as soon as possible

Close monitoring in PICU.
- if continues to experience improvement of GCS, progress towards de-escalation
- if experience any neurological decline, obtain imaging. May need to consider surgery, ICP monitor, and ICP pathway. Also consider EEG.

Elevated ICP, Tier 1 Therapy
- CSF drainage if has EVD
- Hypertonic therapy (3% saline- 1-3ml/kg, 23.4% saline- 0.5ml/kg, manitol-0.5ml/kg).
- Additional analgesia/sedation
- Neuromuscular blockade

If ICP continues elevated and refractory to 1st Tier Therapy, repeat imaging, determine if a surgical option is present.

Tier 2 Therapy
- consider decompressive hemicraniectomy
- consider barbiturate infusion/burst supression
- moderate hypothermia
- hyperventilation (PaCO2 28-34 mmHg)
- higher levels of osmolar therapy (avoid Na >160mEq/L)

Other Considerations:
- if severe TBI, consider obtaining MRI approximately 7-10 days after injury.

**Fig. 1** Algorithm of traumatic brain injury diagnosis and management

c. Imaging typically includes CT scan of the head, cervical spine, chest, abdomen, and pelvis and, depending on the mechanism, CT angiography of the neck and head.
d. Labs obtained during the initial assessment will be utilized to assess the patients' $PaCO_2$, electrolytes, and coagulation profile when determining intracranial pressure effects and possible surgical interventions.
e. Relevant pathologies:
    i. Epidural hematoma (Fig. 2, A) – hematoma in the epidural space, most frequently secondary to a skull fracture, commonly arterial source (MMA) but also from bone diploic vessels or venous sinus injury. Classic lucid interval after the injury before cognitive decline. CTH shows a typical hemorrhage pattern (convex and limited by cranial sutures). Because of the frequent arterial nature of the hemorrhage, can be very large and need prompt surgical intervention for craniotomy and evacuation and hemostasis of bleeding source. Criteria for surgery typically include a hematoma >1 cm in diameter, midline shift >0.5 cm, a neurological deficit, a low GCS, or a decline in GCS.
    ii. Subdural hematoma (Fig. 2, B) – hemorrhage in the subdural space between the arachnoid and dura, typically a venous source such as tearing of a bridging vein or damage to a cortical vein. Acute subdural hematomas are relatively common after trauma and are typically thin with minimal brain compression. They can however be large with similar surgical indications as epidural hematomas. Chronic subdural hema-

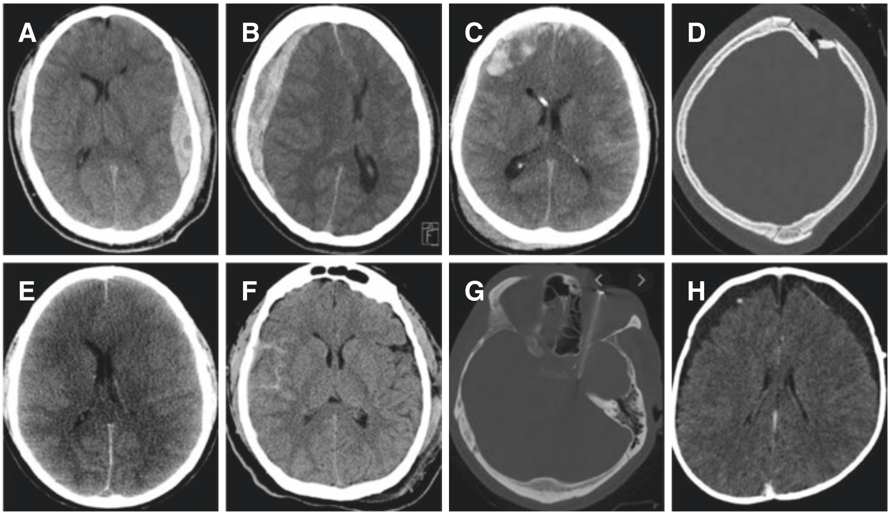


**Fig. 2** CT scans demonstrating pathology detailed below. (A) Left epidural hematoma. (B) Right subdural hematoma. (C) Right frontal contusion. (D) Left frontal depressed skull fracture. (E) Diffuse axonal injury. (F) Right temporal traumatic subarachnoid hemorrhage. (G) Penetrating nail. (H) Bilateral subdural hygromas

tomas are often found in patients with non-accidental trauma and are frequently bilateral. They can be large and exert significant mass effect, although may not have midline shift because of the bilateral forces being applied. Surgical drainage is indicated if the patient is experiencing a neurological deficit or cognitive decline.

iii. Brain contusion (Fig. 2, C) – hemorrhage within the brain tissue (cortex with frequent extension into white matter), damages cortex and fiber tracts and frequently has a moderate amount of local mass effect, but typically without significant mass effect or herniation. Typically non-surgical, but does have a higher likelihood of progressing/expanding within the first 48 h than most other hemorrhages.

iv. Skull fractures (Fig. 2, D) – multiple types of fractures based on type/force of impact and cranial location of the impact.

    1. Non-depressed – can be linear or comminuted, but bone fragments retain their position along the curvature of the skull. Primarily non-operative.
    2. Depressed – fracture of the skull with fragments displaced internally, sometimes protruding through or causing pressure on the brain parenchyma. Surgical repair should be considered (not clearly in pediatric, refer to larger than bone width and talk about babies), if experiencing seizures traceable to the fracture/cortical irritation, if fracture is impinging on important structures, or if concern for cosmetic deformity.
    3. Fractures classified as open or closed (skin can have a laceration, but if the galea is intact, not an open fracture). Depending on the other fracture characteristics, closed fractures can be allowed to heal without surgery. Open fractures are surgical.
    4. Fractures crossing a venous sinus are associated with a risk of venous sinus thrombosis, potentially causing venous congestion that can result in intracranial hypertension and venous strokes. If a patient experiences signs/symptoms concerning for elevated ICP or cerebral ischemia, should obtain vascular imaging (MRV or CTV) to help determine if anticoagulation needs to be initiated [10].
    5. A late consequence of skull fractures is the possibility of developing growing skull fractures, the failure of the fracture to heal, and development of a myelocele or meningocele protruding through the fracture. These occur in children typically less than 5 years old with a skull fracture that involves a dural tear or brain contusion, a cephalohematoma, and bone diastasis of 4 mm or greater [11, 12].
    6. Craniofacial fractures involving the frontal sinus are categorized by the degree of involvement of the anterior and posterior tables.

        a. Treatment involves restoration of anatomy, maintaining a barrier between the intracranial space and the nasopharynx, and preventing varicose complications such as CSF leak, meningitis, etc. [13].

    b. Surgical treatment of frontal sinus fractures depends on fracture pattern, presence of CSF leak, and involvement of the nasofrontal duct [13].
    c. Surgical intervention is recommended for patient with sole anterior table fractures only if there is significant contour deformity and those with open and grossly contaminated fractures regardless of the pattern [13].
    d. Conservative management is recommended for patients with sole posterior table fractures without evidence of nasofrontal duct involvement or CSF leak regardless of posterior table displacement and those with closed fractures of both anterior and posterior table that present with CSF leak.
        i. If patients fail conservative management after 4–7 days, surgical treatment and cranialization are recommended [13]

v. Diffuse axonal injury (DAI) (Fig. 2, E) – severe injury consisting of sheering of axons throughout the white matter, commonly associated with high-speed rotational injury. CT scans may have some small petechial hyperdense areas within the white matter, but frequently they are non-diagnostic. Patients typically have low GCS and neurological exam warranting intracranial pressure monitoring. DAI is better diagnosed on MRI later in the hospital course. Axonal injury and micro-hemorrhage and ischemic damage typically show up on T2, gradient echo, and diffusion-weighted imaging within a week; hence, we target obtaining an MRI around 5–7 days after the injury. A grading system using MRI was established in 1994 assigning a number based on the extent of the injury, extrapolated from a scale determined by histologic slides. Grade 1 includes injury lesions in the hemispheres, essentially at the gray-white junction where shearing forces are highest. Grade 2 involves lesions within the corpus callosum in addition to the hemispheres, and grade 3 deals with lesions in the brainstem plus the corpus callosum and hemispheres. Studies have shown an unfavorable outcome in 17% of grade 1 patients, 40% for grade 2, and 62% for grade 3 [14, 15].

vi. Traumatic subarachnoid hemorrhage (Fig. 2, F) – hemorrhage between the brain and the arachnoid covering from damage to a cortical vessel. Typically small without significant mass effect, not requiring surgery.

vii. Penetrating injuries (Fig. 2, G) – from multiple sources including bullets, dog bites, branches, nails, posts, arrows, knives, and others. Depending on the type and location of the injury, will likely require surgical removal of the foreign bodies, in addition to hematoma evacuations, skull reconstruction, blood vessel repair, or scalp repair.

viii. Non-accidental trauma (Fig. 2, H) – including shaking, blunt force trauma, and penetrating trauma. Infants frequently present with chronic subdural hygromas with signs and symptoms of elevated intracranial pressure manifested by enlarging head circumference, bulging fontanelle, lethargy, and poor feeding. Symptomatic subdural hygromas require drainage with frequent subdural tapping or long-term drainage with a subdural-peritoneal or subdural-subgaleal shunt [16, 17].

ix. Vascular injuries – various vascular injuries associated with trauma include arterial dissections, arteriovenous fistulas, and sinus thromboses and are diagnosed with vascular imaging (CTA/CTV/MRA/MRV/digital subtraction angiography). These types of injuries are frequently asymptomatic at presentation, but patients are at risk for strokes, hemorrhage, and elevated intracranial pressure. Arterial dissection treatment includes aspirin and possible endovascular stenting. Fistulas frequently require endovascular or open surgical repair. Sinus thrombosis is typically treated with anticoagulation and occasionally endovascular thrombectomy.

## Additional Notes

In addition to the immediate surgical treatment for certain pathologies, the algorithm in Fig. 1 has a main focus of preventing elevated ICP while maintaining appropriate cerebral blood flow and oxygenation to avoid secondary brain injury. In patients with severe traumatic brain injury as defined by a GCS less than 9 with imaging findings of injury, an ICP monitor is placed to be able to track changes and elevations [18–20]. ICP management is typically divided into basic maintenance therapies followed by more aggressive tier one and tier two levels of therapy as detailed in the algorithm [21, 22].

## Study Questions

1. A 10-year-old patient is brought to the emergency department after crashing on her bike without a helmet. Her scalp is bleeding, she is not opening her eyes to voice or stimulation, she is not talking or making any verbal sounds, but she is localizing with her right arm. Her left pupil is 3 mm and reactive; her right pupil is 5 mm and unreactive. What is the first step in management after she arrives in the trauma bay?

   a. Obtain an emergent CT head.
   b. Call neurosurgery for an emergent ICP monitor placement.
   c. Perform ATLS/PALS.
   d. Give 1 mg/kg of mannitol.

   Answer: (c) ATLS/PALS pathways always take precedence prior to more focused injury assessment and diagnosis.

2. What is the recommended initial management for a closed fracture of the frontal sinus with a CSF leak?

   a. CSF diversion
   b. Conservative management with observation
   c. Craniotomy for repair of dural defect
   d. Cisternogram

   Answer: (b) Conservative management with observation. The vast majority of closed frontal sinus fractures with CSF leak will resolve spontaneously. Initial management is observation. If the CSF leak persists for 4–7 days after observation, more invasive options can be explored.

3. With the same patient, what is her GCS?

   a. 11
   b. 7
   c. 8
   d. 6

   Answer: (b) Her GCS is 7 with one point for no eye opening, one point for no verbal sounds, and five points for localizing with her right upper extremity.

4. The patient is intubated and stabilized. A CT scan of her head shows a 1 cm × 5 cm right temporal epidural hematoma with 9 mm of midline shift. What is the next step in management?

   a. Place an emergent external ventricular drain.
   b. Transfer to PICU for monitoring.
   c. Give hyperosmolar therapy and take for emergent surgical evacuation.
   d. Obtain an emergent MRI of her brain.

   Answer: (c) This patient has been stabilized, and the acute epidural hematoma has been identified on the trauma scans with significant mass effect with midline shift. She needs prompt surgical drainage to minimize secondary injury. Giving hyperosmolar therapy immediately can help reduce the ICP while transitioning to the operating room.

5. The patient undergoes a craniotomy and epidural hematoma evacuation with an intraoperative external ventricular drain. Four days later, her ICP is noted to persistently be around 25 mmHg. What are appropriate tier 1 steps to take to control her ICP?

   a. Lay the patient flat and monitor; give the patient a bolus of normal saline to increase the MAP to maintain CPP.
   b. Ensure the head of bed is 30° and straight; check $PaCO_2$, sedation level, and temperature.
   c. Sit patient up to 60°; decrease her sedation to obtain a better neuroexam.
   d. Drop the EVD to 0 mmHg, and drain 25 ml of CSF

Answer: (b) According to the Brain Trauma Foundation guidelines, maintaining the head of bed at 30° and straight provides for better venous outflow; the $PaCO_2$ should be checked to make sure she is not hypercarbic, her temperature should be checked to make sure she is not hyperthermic, and she should be adequately sedated. All of these aspects have significant impact on ICP if not in the appropriate range and are typically correctable in short order.

## References

1. Kochanek PM, Tasker RC, Bell MJ, Adelson PD, Carney N, Vavilala MS, Selden NR, Bratton SL, Grant GA, Kissoon N, Reuter-Rice KE, Wainwright MS. Management of pediatric severe traumatic brain injury: 2019 consensus and guidelines-based algorithm for first and second tier therapies. Pediatr Crit Care Med. 2019a Mar;20(3):269–79.
2. Kochanek PM, Tasker RC, Carney N, Totten AM, Adelson PD, Selden NR, Davis-O'Reilly C, Hart EL, Bell MJ, Bratton SL, Grant GA, Kissoon N, Reuter-Rice KE, Vavilala MS, Wainwright MS. Guidelines for the management of pediatric severe traumatic brain injury, third edition: update of the brain trauma foundation guidelines. Pediatr Crit Care Med. 2019b;20:S1–S82.
3. Carney N, Totten AM, O'Reilly C, et al. Guidelines for the management of severe traumatic brain injury, fourth edition. Neurosurgery. 2017;80:6–15.
4. Appavu B, Foldes ST, Adelson PD. Clinical trials for pediatric traumatic brain injury: definition of insanity? J Neurosurg Pediatr. 2019;23(6):661–9.
5. Langlois JA, Rutland-Brown W, Thomas KE. The incidence of traumatic brain injury among children in the United States: differences by race. J Head Trauma Rehabil. 2005;20(3):229–38.
6. Taylor CA, Bell JM, Breiding MJ, Xu L. Traumatic brain injury-related emergency department visits, hospitalizations, and deaths—United States, 2007 and 2013. MMWR Surveill Summ. 2017;66(9):1–16.
7. Jha RM, Kochanek PM. Adding insight to injury: a new era in neurotrauma. Lancet Neurol. 2017;16:578–80.
8. DeKosky ST, Asken BM. Injury cascades in TBI-related neurodegeneration. Brain Inj. 2017;31:1177–82.
9. Kochanek PM, Jackson TC, Ferguson NM, et al. Emerging therapies in traumatic brain injury. Semin Neurol. 2015;35:83–100.
10. Hersh DS, Shimony N, Groves ML, Tuite GF, Jallo GI, Liu A, Garzon-Muvdi T, Huisman TAGM, Felling RJ, Kufera JA, Ahn ES. Pediatric cerebral venous sinus thrombosis or compression in the setting of skull fractures from blunt head trauma. J Neurosurg Pediatr. 2018 Mar;21(3):258–69.
11. Singh I, Rohilla S, Siddiqui SA, Kumar P. Growing skull fractures: guidelines for early diagnosis and surgical management. Childs Nerv Syst. 2016;32(6):1117–22.
12. Le Fournier L, Hénaux PL, Haegelen C, Proisy M, Riffaud L. Intradiploic growing skull fracture: review of mechanisms and literature. Childs Nerv Syst. 2015;31(11):2199–205. https://doi.org/10.1007/s00381-015-2793-4.
13. Vu AT, Patel PA, Chen W, Wilkening MW, Gordon CB. Pediatric frontal sinus fractures: outcomes and treatment algorithm. J Craniofac Surg. 2015;26(3):776–81.
14. van Eijck MM, Herklots MW, Peluso J, Schoonman GG, Oldenbeuving AW, de Vries J, van der Naalt J, Roks G. Accuracy in prediction of long-term functional outcome in patients with traumatic axonal injury: a comparison of MRI scales. Brain Inj. 2020;34(5):595–601.
15. Gentry LR. Imaging of closed head injury. Radiology. 1994;191(1):1–17.
16. Korinth MC, Lippitz B, Mayfrank L, Gilsbach JM. Subdural-atrial and subdural-peritoneal shunting in infants with chronic subdural fluid collections. J Pediatr Surg. 2000;35(9):1339–43.

17. Blauwblomme T, Di Rocco F, Bourgeois M, Beccaria K, Paternoster G, Verchere-Montmayeur J, Sainte-Rose C, Zerah M, Puget S. Subdural to subgaleal shunts: alternative treatment in infants with nonaccidental traumatic brain injury? J Neurosurg Pediatr PED. 2015;15(3):306–9.
18. Alkhoury F, Kyriakides TC. Intracranial pressure monitoring in children with severe traumatic brain injury: National trauma data bank-based review of outcomes. JAMA Surg. 2014;149:544–8.
19. Bennett TD, DeWitt PE, Greene TH, et al. Functional outcome after intracranial pressure monitoring for children with severe traumatic brain injury. JAMA Pediatr. 2017;171:965–71.
20. Stippler M, Ortiz V, Adelson PD, et al. Brain tissue oxygen monitoring after severe traumatic brain injury in children: relationship to outcome and association with other clinical parameters. J Neurosurg Pediatr. 2012;10:383–91.
21. Shein SL, Ferguson NM, Kochanek PM, Bayir H, Clark RS, Fink EL, Tyler-Kabara EC, Wisniewski SR, Tian Y, Balasubramani GK, Bell MJ. Effectiveness of pharmacological therapies for intracranial hypertension in children with severe traumatic brain injury—results from an automated data collection system time-synched to drug administration. Pediatr Crit Care Med. 2016;17(3):236–45.
22. Bell MJ, Adelson PD, Hutchison JS, et al. Multiple medical therapies for pediatric traumatic brain injury workgroup: differences in medical therapy goals for children with severe traumatic brain injury-an international study. Pediatr Crit Care Med. 2013;14:811–8.

## *Further Reading*

Appavu B, Foldes ST, Adelson PD. Clinical trials for pediatric traumatic brain injury: definition of insanity? J Neurosurg Pediatr. 2019;23(6):661–9.

Kochanek PM, Tasker RC, Bell MJ, Adelson PD, Carney N, Vavilala MS, Selden NR, Bratton SL, Grant GA, Kissoon N, Reuter-Rice KE, Wainwright MS. Management of pediatric severe traumatic brain injury: 2019 consensus and guidelines-based algorithm for first and second tier therapies. Pediatr Crit Care Med. 2019a Mar;20(3):269–79.

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# Abdominal Trauma

**James DeCou, Sarah Alander, and Christopher P. Coppola**

**Key Points**

1) Management of abdominal trauma in children should follow the guidelines of Advanced Trauma Life Support (ATLS) with systematic evaluation of airway, breathing, circulation, disability, and exposure (ABCDE's) followed by a head-to-toe secondary survey.
2) The majority of abdominal trauma in children is blunt, with only about 10% of abdominal trauma in children due to penetrating etiology.
3) The vast majority of blunt abdominal trauma can be managed non-operatively, the exceptions being hemorrhagic shock due to solid organ injury and perforation of hollow viscus.
4) Nearly all penetrating abdominal injury in children will require operative treatment.
5) When possible, the spleen should be preserved in injured children to avoid a vulnerability to overwhelming post-splenic infection (OPSI).

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1. Blunt injury
   a. Blunt force is the most common mechanism of trauma in children [1].
   b. Attention should be given to the degree of energy involved, such as in the case of a baseball bat injury versus a high-speed motor vehicle collision. The injury mechanism and history predict injury pattern and aid in choosing highest yield diagnostic procedures.
2. Penetrating injury
   a. Penetrating abdominal injuries are most commonly the result of firearm or stabbing injuries [2].
   b. Significant intraperitoneal injuries are present in most children who sustain gunshot wounds, and, as a result, abdominal exploration is usually needed. Likewise, stab wounds to the abdomen will almost always require explorative laparotomy.
3. Specific organ injury
   a. Spleen
      i. The spleen is the most commonly injured abdominal organ in children, whereas the liver is most commonly injured in adults.
      ii. The standard of care in treating hemodynamically stable children with splenic injury is observation alone. Non-operative management is successful in greater than 90% of children with splenic injury.
      iii. Indications for operative intervention are generally limited to persistent hypotension, greater than 50% blood volume replacement, or additional life-threatening injuries.
      iv. Intravenous access should be obtained via a large-bore cannula, and the patient should be on bed rest initially, and for high-grade injuries, monitored in the pediatric intensive care unit.
      v. In addition to routine cardiorespiratory monitoring, frequent hemoglobin measurements should be obtained over the initial 24–48 h to assess for ongoing hemorrhage. Noninvasive hemoglobin monitoring is an emerging technology that may decrease the number of blood draws needed for lower-grade injuries.
   b. Liver
      i. The liver is the second most injured abdominal organ in children. Like splenic injury, liver injuries can be managed non-operatively in most hemodynamically stable children.
      ii. Ecchymosis or abrasion over the right upper quadrant may suggest significant injury.
      iii. Complications of liver injury include hemorrhage, bile peritonitis, abscess, and hemobilia.
      iv. Operative intervention is typically required in the case of disruption of the hepatic veins or retrohepatic caval injuries.

    v. Hepatic vessel embolization is sometimes utilized for select patients, particularly those who have ongoing hemorrhage and a blush of contrast seen on computed tomography.
    vi. Significant bile leak may require interventional radiology drain placement and/or endoscopic retrograde cholangiopancreatography (ERCP) with stent placement.

c. Pancreas

    i. Injuries

        1. Pancreatic injuries may require operative intervention, especially if the pancreatic duct is transected.
        2. Transection of the pancreas usually occurs where the gland crosses the spine.
        3. Late complication after injury is development of pseudocyst.

    ii. History may suggest pancreas injury

        1. Seatbelt injury, particularly when only lap belt is used and is positioned too high above the iliac crests.
        2. Handlebar injury, where a child falls forward onto a handlebar during a bicycle crash.
        3. Punch or kick to the epigastrium.
        4. A “seat belt sign” or “handlebar sign” with a bruise over the mid abdomen should raise suspicion for a pancreas injury.

    iii. Clinical signs

        1. Upper abdominal seat belt sign, handlebar contusion, and/or tenderness that may radiate to back
        2. Elevated amylase and lipase levels
        3. Computed tomography revealing gland edema, fluid in lesser sac, transaction, or hemorrhage of gland

    iv. Treatment

        1. Transaction of the pancreas is best treated by early distal pancreatectomy with drainage.
        2. If pancreatic duct is intact, non-operative treatment with bowel rest and total parenteral nutrition is likely best treatment.
        3. ERCP may be useful in diagnosing duct disruption and in treating late complications of stricture and fistula.
        4. Pseudocyst, if it occurs, should be allowed 6 weeks for the lining to mature and then is drained into the GI tract. Options include endoscopic transgastric drainage and laparoscopic Roux-en-Y pseudocyst-jejunostomy.

d. Kidney

   i. Injury

      1. Blunt trauma to the abdomen or flank can result in laceration, hematoma, or vascular injury.
      2. Penetrating injury to kidney can cause laceration, hemorrhage, and vascular injury.
      3. Late effects can be abscess, urinoma, or scar causing hypertension.

   ii. Clinical signs

      1. Flank tenderness, mass, pain, and ecchymosis.
      2. Gross or microscopic hematuria, although it is not present in all renal injuries.
      3. Sonography will detect renal laceration, hematoma, and fluid collections.
      4. Computed tomography can diagnose and grade renal injury.

   iii. Treatment

      1. Non-operative management will usually be successful for renal trauma if patient remains hemodynamically stable.
      2. Ongoing hemorrhage from a kidney rarely may require open exploration.
      3. Renal salvage can be attempted, but often nephrectomy is the safest choice.
      4. Before removing a kidney, the surgeon must verify that the opposite kidney is present.
      5. If available and patient is stable enough to try the procedure, angiographic embolization can be attempted.
      6. Patients with urinary extravasation receive antibiotics.
      7. Late intervention may be needed for abscess, infected urinoma, or renovascular hypertension.

e. Duodenum

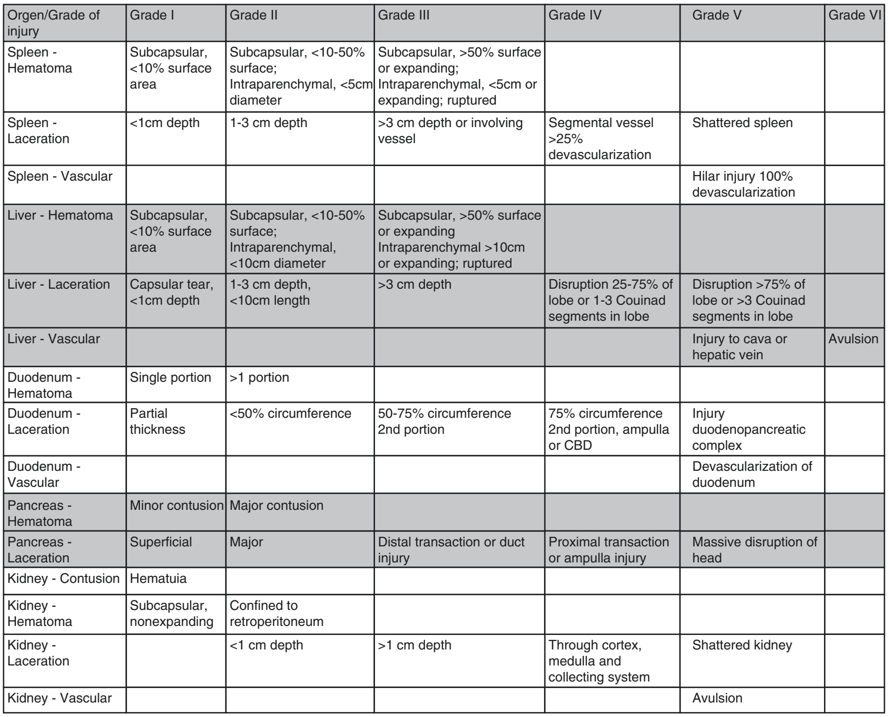
   i. Duodenal injuries are uncommon; however, they have a higher rate of occurrence than the adult trauma population.
   ii. Most duodenal injuries present as duodenal hematoma without intraluminal perforation. Thickening of the duodenal wall is noted on computed tomography scan. These injuries are treated with observation and parenteral nutrition. For select patients, a nasojejunal feeding tube will allow post-duodenal enteral feeding.
   iii. Full-thickness perforation will typically present with extravasation of oral contrast in the right anterior pararenal space. Most of these injuries are amenable to debridement and primary closure; however more complex approaches are sometimes needed.

f. Jejunum, ileum, colon

  i. Bowel injuries can frequently be difficult to diagnose. A high index of suspicion with serial abdominal exams is often required to make the diagnosis.
  ii. Suspicion is raised in patients with free intra-abdominal fluid without solid organ injury on CT.
  iii. In patients with bowel injuries, studies have shown that morbidity does not increase if the injury is diagnosed and surgically treated within 24 h of injury.
  iv. Laparoscopy has been used to evaluate possible bowel injuries with both blunt and penetrating mechanisms.

4. Scales for grading organ injury (Fig. 1) [3]
5. Common patterns of injury

  a. Seat belt trauma



| Orgen/Grade of injury | Grade I | Grade II | Grade III | Grade IV | Grade V | Grade VI |
|---|---|---|---|---|---|---|
| Spleen - Hematoma | Subcapsular, <10% surface area | Subcapsular, <10-50% surface;<br>Intraparenchymal, <5cm diameter | Subcapsular, >50% surface or expanding;<br>Intraparenchymal, <5cm or expanding; ruptured | | | |
| Spleen - Laceration | <1cm depth | 1-3 cm depth | >3 cm depth or involving vessel | Segmental vessel >25% devascularization | Shattered spleen | |
| Spleen - Vascular | | | | | Hilar injury 100% devascularization | |
| Liver - Hematoma | Subcapsular, <10% surface area | Subcapsular, <10-50% surface;<br>Intraparenchymal, <10cm diameter | Subcapsular, >50% surface or expanding<br>Intraparenchymal >10cm or expanding; ruptured | | | |
| Liver - Laceration | Capsular tear, <1cm depth | 1-3 cm depth, <10cm length | >3 cm depth | Disruption 25-75% of lobe or 1-3 Couinad segments in lobe | Disruption >75% of lobe or >3 Couinad segments in lobe | |
| Liver - Vascular | | | | | Injury to cava or hepatic vein | Avulsion |
| Duodenum - Hematoma | Single portion | >1 portion | | | | |
| Duodenum - Laceration | Partial thickness | <50% circumference | 50-75% circumference 2nd portion | 75% circumference 2nd portion, ampulla or CBD | Injury duodenopancreatic complex | |
| Duodenum - Vascular | | | | | Devascularization of duodenum | |
| Pancreas - Hematoma | Minor contusion | Major contusion | | | | |
| Pancreas - Laceration | Superficial | Major | Distal transaction or duct injury | Proximal transaction or ampulla injury | Massive disruption of head | |
| Kidney - Contusion | Hematuia | | | | | |
| Kidney - Hematoma | Subcapsular, nonexpanding | Confined to retroperitoneum | | | | |
| Kidney - Laceration | | <1 cm depth | >1 cm depth | Through cortex, medulla and collecting system | Shattered kidney | |
| Kidney - Vascular | | | | | Avulsion | |

**Fig. 1** Organ injury grading scale. (Source of information in Table: Moore EE, MD, Cogbill TH, Malangoni M, Jurkovich GJ, Champion HR. Scaling system for organ specific injuries. The American Association for the Surgery of Trauma. http://www.aast.org/Assets/56ef079d-229c-45f2-9b18-c3825e450e65/633867256925730000/injuryscoringtables-pdf. Accessed 20 Oct 2021)

    i. Seat belts must contact the body in the proper location. Children less than 8 years old or 4 feet 9 inches tall need a booster suit so that the lap belt crosses over the anterior superior iliac crests.
    ii. When a lap belt is used either without a shoulder strap or without a booster seat when appropriate, it can result in injury.
    iii. In a head-on motor vehicle crash, if a child is using a lap belt only, the body is forcefully flexed at the waist.
    iv. The lap belt crushes the viscera against the spine.
    v. Organs at risk are the colon, small bowel, mesentery, duodenum, pancreas, and aorta.
    vi. Hyperflexion of the spine can result in a Chance fracture: transverse distraction fracture of the posterior portion of the vertebral body.

b. Handlebar injury

    i. In a bike crash, if the handlebars turn acutely, the child can receive a direct blow to the abdomen with the end of the handlebar, often leaving a round contusion on the abdominal wall.
    ii. This can cause injury to pancreas, perforation of a hollow viscus, or traumatic hernia.

c. Waddell's triad

    i. Constellation of injuries when a pedestrian is struck by a car.
    ii. The components of the triad are femur or tibial fracture, intra-abdominal or intra-thoracic injury, and subdural hematoma, but not all three injuries are always present [4].
    iii. The bumper of the car strikes the child's leg and causes a fracture of the femur or the tibia and fibula, depending on the height of the bumper relative to the child. (In countries where cars drive on the right, the child is usually struck on the left side. In countries where cars drive on the left side of the road, it is the opposite.)
    iv. When the child is thrown up on the hood of the vehicle, intra-abdominal injury (e.g., spleen, liver, bowel) or intrathoracic injury (e.g., pulmonary contusion) occurs.
    v. When the child's head strikes the windshield or the vehicle brakes throwing the child to the ground, subdural hematoma results.

d. Non-accidental trauma (NAT)

    i. Head injury and fractures are most common injuries in NAT, but abdominal injury is second most common fatal injury [5].
    ii. Children with NAT abdominal injury tend to the younger (0–36-month-old) than children with abdominal injury from accidental trauma [6].
    iii. Bowel injury is disproportionately increased in NAT; the liver and pancreas are the most frequently injured organs.
    iv. Children with NAT abdominal injury are less likely to have bruising or distension. For this reason, testing of transaminase for children younger

than age 5 years old is recommended for NAT evaluations. Aspartate aminotransferase (AST) or alanine aminotransferase (ALT) levels higher than 80 IU/L prompt evaluation with contrast CT [7].

6. Assessment
    a. Initial assessment follows Advanced Trauma Life Support assessment with stabilization of C-spine and evaluation of airway, breathing, and circulation.
    b. Physical examination: Abdomen is examined for ecchymosis, laceration, distension, masses, and tenderness.
    c. FAST
        i. Focused Abdominal Sonography for Trauma (FAST) is a rapid, noninvasive, and portable method to evaluate the abdomen.
        ii. Sonographic windows examined for abnormal fluid collections are the pericardium, Morrison's pouch between liver and right kidney, the space between spleen and left kidney, and the retrovesical space.
        iii. Right and left pleural spaces are evaluated for pneumothorax, indicated by the absence of normal lung sliding.
        iv. The finding of free fluid in abdomen on FAST suggests solid organ injury and need for more diagnostic maneuvers (i.e., CT scan).
    d. Peritoneal lavage
        i. Used much less frequency since FAST became available.
        ii. Peritoneal cavity is entered via a small supraumbilical incision.
        iii. Abdominal fluid is sampled for blood, amylase, or intestinal contents.
        iv. Can diagnose injury to solid organ or perforated viscus.
        v. Will not diagnose retroperitoneal injury with contained bleeding.
    e. CT scan
        i. Requires ionizing radiation with its associated cumulative risk of malignancy.
        ii. Ensure that patient is hemodynamically stable before risking a trip to the scanner.
        iii. Can diagnose and grade injury to solid organs.
        iv. Able to diagnose free fluid and air from hollow viscus injury with 94% sensitivity, but signs of perforation may require time to develop after injury.
    f. MRI
        i. Used less frequently to evaluate trauma.
        ii. Requires more time for images and may require anesthetic for child to stay still.
        iii. Can be used to evaluate for injury to the biliary tree through magnetic resonance cholangiopancreatography (MRCP).

g. ERCP

    i. Useful in evaluating and treating pancreas injury and bile leak from liver injury.
    ii. Endoscopic ultrasound can be performed at the same time.
    iii. If a pancreatic duct injury is detected, a stent can be placed.
    iv. If a significant bile leak is noted, common bile stenting is useful.
    v. Limited availability in most pediatric trauma facilities especially for younger patients.

h. Angiography

    i. Angiography with embolization offers an intermediate step for patients with hemorrhage from solid organ injury.
    ii. Embolization can be performed to the liver, spleen, kidney, and pelvic vessels.
    iii. Angiography carries the risk of vascular injury and hemorrhage.
    iv. Indications for embolization vary, but may include blush on computed tomography, ongoing hemorrhage and blood transfusion requirement, and a patient stable enough to undergo the procedure.

i. Intravenous pyelography (IVP)

    i. For suspected kidney, ureter, or bladder injury, IVP can help define anatomy if injury.
    ii. This data may be already present if CT scan included delayed views of renal contrast excretion.

7. Treatment

    a. Correction of shock with intravenous boluses of crystalloid fluid and transfusion of blood for non-responders.
    b. Non-operative treatment

        i. Most solid organ injuries can be managed non-operatively.
        ii. Patients are admitted and are monitored for vital signs and hemoglobin at regular intervals.
        iii. Patients are initially kept NPO for possible laparotomy.
        iv. Criteria are established for failure of non-operative therapy such as hemodynamically unstable patient or transfusion requirement greater than 6 units of blood or half a patient's blood volume.
        v. Hemodynamically stable patients can rapidly be progressed to oral diet, ambulation, and transfer to regular patient ward if they were admitted to the Pediatric Intensive Care Unit upon admission.
        vi. Patients with solid organ injury will need restriction from sports and rough activity for 3–6 weeks after injury, depending on the grade of injury, to facilitate recovery without recurrence of bleeding [8] (Fig. 2).

| CT grade | I | II | III | IV |
|---|---|---|---|---|
| ICU stay (days) | None | None | None | 1 |
| Hospital stay (days) | 2 | 3 | 4 | 5 |
| Pre-discharge imaging | None | None | None | None |
| Post-discharge imaging | None | None | None | None |
| Activity restriction (weeks)[a] | 3 | 4 | 5 | 6 |

**Fig. 2** American Pediatric Surgical Association recommendations for isolated liver or spleen injury CT grade. *Return to full-contact, competitive sports (i.e., football, wrestling, hockey, lacrosse, and mountain climbing) should be at the discretion of the individual pediatric trauma surgeon. The proposed guidelines for return to unrestricted activity include "normal" age-appropriate activities (Adapted from ref. [8])

c. Operative treatment

  i. Damage control surgery

    1. Hemodynamically unstable patients who do not respond to volume resuscitation undergo emergent laparotomy.
    2. Wide exposure with a midline laparotomy is obtained, and sponges are packed into all four quadrants and the pelvis.
    3. Volume resuscitation continues through the procedure.
    4. Each quadrant is unpacked one at a time, and efforts are made to diagnose all injuries and control bleeding.
    5. Bowel injuries can be temporarily stapled off, with plans for definitive repair at a second-look laparotomy.
    6. If bleeding cannot be controlled quickly, the abdomen is left packed with sponges with temporary abdominal closure.
    7. The patient is brought to the pediatric intensive care unit (PICU) for rewarming, continued resuscitation, and correction of abnormalities of coagulation.
    8. The patient returns to the OR for washout of abdomen in 24–48 h with more definitive correction of injuries.

  ii. Decompressive laparotomy

    1. Hemorrhage and edema after abdominal trauma can increase the pressure within the peritoneal cavity.
    2. Increased pressure will result in abdominal compartment syndrome with reduction in urine output, reduced return of blood to heart, and compromised blood flow to lower extremities. It has also been shown to worsen intracranial pressure in patients with traumatic brain injuries.
    3. Pressure is measured via a Foley catheter in the bladder. Pressure is normally 12 mm Hg or less, and laparotomy must be considered for pressure above 25 mm Hg.

    4. Decompressive laparotomy is wide midline laparotomy allowing the edematous viscera and retroperitoneum to bulge out and temporary abdominal closure.
    5. Abdomen is washed out every 2–3 days, and fascia is slowly closed as edema decreases.

iii. Temporary abdominal closure and vacuum-assisted closure (VAC): VAC can be used as a temporary dressing when the abdomen is left open after laparotomy. The continuous suction removes fluid collections which could result in bacterial overgrowth and abscess. It also reduced edema fluid of viscera.

iv. Procedures for liver

    1. Most injuries can be treated non-operatively.
    2. If operative intervention is required due to shock or ongoing blood loss, consider damage control surgery with a second look later.
    3. Embolization can be employed before or after laparotomy.
    4. Bleeding from the vena cava or hepatic veins requires operative control of hemorrhage and has a high incidence of morbidity and mortality [9].
    5. Biloma can be drained percutaneously by interventional radiology. ERCP with common bile duct stenting can be useful in managing ongoing bile leak from a severe liver injury.

v. Splenectomy and splenorrhaphy

    1. Operative intervention for splenic trauma usually results in splenectomy.
    2. Attempts can be made at splenic salvage with splenorrhaphy using transfixion suture, omentum, wrapping with mesh, or a combination of techniques.
    3. Patients with splenectomy or major injury to spleen require antibiotic therapy plus vaccination against infection from pneumococcus, *Haemophilus influenzae*, and meningococcus.

vi. Pyloric exclusion (Fig. 3)

    1. Minor injuries of the duodenum can be primarily repaired and drained if there is not shock, major contamination, and viable duodenal tissue.
    2. Major injuries of the duodenum do better with washout, wide drainage, and pylorus exclusion either by stapling across the distal stomach (without division) or open pylorus exclusion by opening stomach and closing lumen of distal stomach with running suture placed from the inside.
    3. Stomach is drained through a gastrojejunostomy and sometimes gastrostomy.

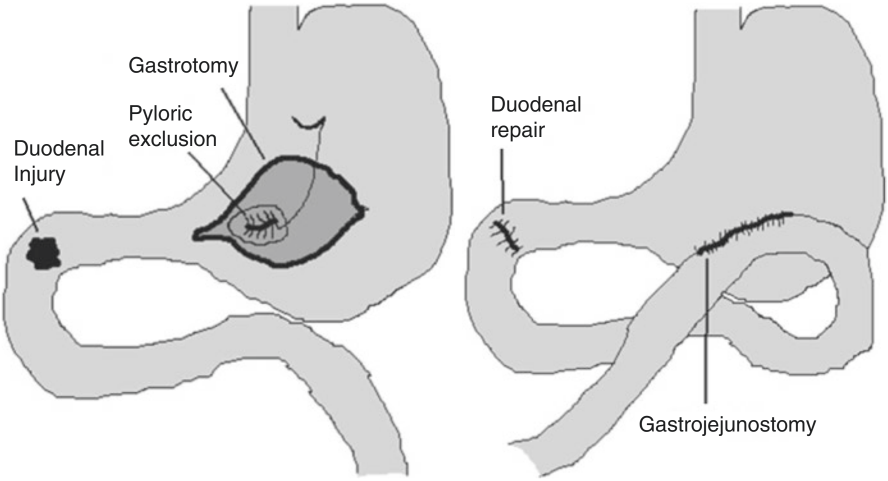


**Fig. 3** Pyloric exclusion. Major trauma to the duodenum is best treated with pyloric exclusion. The stomach is opened, and through gastrotomy, pylorus is closed with running suture. The injury to duodenum is debrided, closed, and widely drained. The stomach is drained with a gastrojejunostomy. (Source: Christopher Coppola)

vii. Exploration of retroperitoneum

   1. Complete abdominal exploration sometimes includes mobilization of retroperitoneal structures via a medial visceral rotation maneuver [10].
   2. Whether stable or expanding, most hematomas of the retroperitoneum should be explored. Nonexpanding hematomas of the perirenal or retrohepatic region are typically not explored, and instead they are treated with packing.

viii. Kocher maneuver

   1. Injuries involving a high degree of suspicion for duodenal or pancreatic injury require mobilization of the duodenum and examination of the retroperitoneum.
   2. The Kocher maneuver involves dissection of the lateral peritoneal attachments of the duodenum to expose all three segments of the duodenum, as well as the pancreatic head and neck.

ix. Medial visceral rotation

   1. Rapid exposure of the major abdominal vessels can be obtained by utilizing mobilization maneuvers, whereas the viscera are rotated off the midline. There are two common approaches: left-sided and right-sided medial visceral rotation [6].
   2. Left-sided medial visceral rotation (Mattox maneuver) will expose the entire abdominal aorta along with its branches, with exception to

the right renal artery. It is carried out by incising the lateral peritoneal attachment (white line of Toldt) about the left colon and sigmoid, then along the splenic flexure, around the posterior spleen, and behind the gastric fundus. The stomach, pancreas, spleen, and left colon are then rotated toward the midline.

3. Right-sided medial visceral rotation (Cattell-Braasch maneuver) is utilized when access is needed to the infrahepatic inferior vena cava. The right colon is mobilized by taking down the hepatic flexure of the colon and then continuing dissection along the paracolic gutter via the peritoneal reflection. Further blunt dissection will allow the colon to be rotated medially toward the aorta. If additional exposure of the vena cava is needed below the liver, then the Kocher maneuver may be utilized with medial mobilization of the duodenum and pancreatic head.

x. Nephrectomy and ureter repair

1. Exploration is warranted in children who are hemodynamically unstable, have an expanding hematoma, or have injury to other nearby viscera warranting exploration.
2. Presences of a contralateral kidney should be confirmed prior to proceeding with nephrectomy. This fact may be known through prior CT scan if the child was stable enough to get one before operation. Nephrectomy is the safest choice for children with unstable injuries. Delayed nephrectomy may be warranted in the patients who develop hypertension or symptomatic renal infarction.
3. Ureteral repair may require a staged procedure in patients who are too unstable. For staged repair, the damaged ureter is commonly tied off with long silk ties to assist in localization later. Children who exhibit shock are at higher risk of complications due to tissue hypoxia. These complications include stricture, urinoma, fistula, and loss of tissue.
4. Partial transection can typically be repaired primarily, with exception to gunshot wounds.
5. Injuries of the upper ureter are typically repaired with ureteroureterostomy if a tension-free anastomosis is achieved. Mobilization of the kidney may be required to reduce tension. Ureteropelvic junction injury is more common in children than adults. Injuries involving ureteral avulsion from the renal pelvis may be anastomosed directly into the renal pelvis as an ureteropyelostomy procedure. Ureterocalicostomy may be performed in situations where the renal pelvis or ureteropelvic junction is damaged beyond repair.
6. Mid-ureteral injuries are classically repaired with ureteroureterostomy. In extreme cases where there is extensive loss of ureter, transureteroureterostomy via donor ureter may be utilized.
7. Lower ureteral injuries are often accompanied by disruption of blood supply from the iliac vessels. As a result, these injuries are often

repaired via ureteroneocystostomy, whereas the ureter is re-implanted into the bladder. This repair is typically stented, and a suprapubic tube and urethral Foley catheter are placed. For longer defects, the bladder may be mobilized and hitched to the psoas tendon to bridge the defect. Urethral anastomosis is then carried out medial to the hitch.

8. Consultation with a specialized pediatric urology team can help in management of these injuries, and for some children cystoscopy and stent placement can help with urinary obstruction and leak as well as encourage healing of injury without laparotomy.

xi. Pancreatectomy and trauma Whipple

1. Operative intervention is typically related to the integrity of the pancreatic duct. In cases of blunt trauma, the pancreas is usually fractured where it crosses the vertebral column. Early distal pancreatectomy and drainage is preferred in cases of pancreatic transection. If an intact duct is confirmed, then non-operative management is an acceptable consideration.
2. Penetrating trauma to the head and neck without duct injury can be treated with simple drainage. When the ductal system is compromised, Roux-en-Y pancreaticojejunostomy is sometimes performed.
3. Severe injuries involving devascularization of the pancreas and duodenum require pyloric exclusion, drainage, and repeat debridement. While rare, children with catastrophic injury of the pancreatic head might require trauma Whipple or pancreaticoduodenectomy.

## Additional Notes

Trauma is the most common cause for hospitalization and death in children, so any emergency provider caring for children should be prepared to care for injured children. Trauma care should follow the guidelines of ATLS, and children with serious injuries need to be transferred to a pediatric trauma center for specialized care.

The practice of treating solid organ injuries with non-operative management is one that started at pediatric trauma centers and was later adopted by adult trauma centers. Time has shown that very few children with blunt solid organ injury will need operative treatment, even when the injury is high grade. If the patient remains hemodynamically stable and there is no sign of hollow organ viscus rupture, non-operative treatment should be continued. In contrast, penetrating injury to the abdomen carries higher mortality and most children will need laparotomy to identify the nature of the trauma and repair injuries at sites of viscus rupture [1].

Children who suffer hemorrhagic shock after abdominal trauma will need resuscitation according to the Advanced Trauma Life Support guidelines. If infusion of a crystalloid bolus does not restore perfusion, the child will need transfusion. Some modern developments to assist resuscitation are transfusion of whole blood, rather than packed red blood cells, and massive transfusion protocols with 1:1:1 matched component transfusions of packed red blood cells, plasma, and platelets to avoid development of coagulopathy [11]. Thromboelastography can guide specifically which blood component is in deficit and needs transfusion to restore. The antifibrinolytic agent tranexamic acid (TXA) has been shown to reduce hemorrhage and mortality in severely injured children [12].

In select patients with solid organ injury, catheter-based therapy by an interventional radiologist may help reduce the rate of hemorrhage when an active blush of bleeding is seen on computed tomography [13]. Care must be taken to ensure that the child is stable enough to make it safely through the procedure. Unstable patients are better served with an immediate damage control laparotomy to address active hemorrhage and continue resuscitation. Some institutions have specialized hybrid operating rooms where both open laparotomy and fluoroscopy-guided catheter-based interventions can be performed simultaneously.

## Study Questions

1) Which injury is not typically part of Waddell's triad?

   a. Femur fracture
   b. Thermal injury
   c. Splenic rupture
   d. Subdural hematoma

   Answer to Question 1: (b) Thermal injury. Waddell's triad is a constellation of injuries that occurs when a child is struck by a vehicle. The triad includes femur fracture, intra-abdominal/intrathoracic injury, and subdural hematoma. Thermal injury is not part of Waddell's triad. The leg is injured when the bumper of the vehicle strikes the child's leg. When the child is thrown up on the hood of the car, abdominal or thoracic organs, such as the spleen, are injured. After the car brakes, the child is thrown to the ground and the head striking the pavement results in a subdural hematoma. It is important to note that that this triad of injuries does not occur every time a pedestrian is struck but knowing the history of the trauma helps the provider to actively seek injuries that have a higher probability of being present.

2) How many months should a child who sustains an isolated grade II splenic laceration that is successfully managed non-operatively be restricted from sports and rough activity?

   a. 1 month
   b. 2 months
   c. 3 months
   d. 4 months

Answer to Question 2: (a) 1 month. According to the American Pediatric Surgical Associations recommendations for isolated spleen or liver injury, after a grade II splenic injury, 1-month restriction from sports and rough activity is sufficient recovery. If the child has an asymptomatic recovery, no routinely scheduled follow-up sonographic or computed tomography imaging is necessary. The guidelines note that the specific post-injury recommendations are at the discretion of the individual treating pediatric surgeon.

3) What is the best treatment for an isolated pancreatic injury caused by a handlebar injury in which computed tomography reveals transection of the mid portion of the pancreas gland and pancreatic duct?

    a. Non-operative monitoring and feeding trial
    b. Whipple pancreaticoduodenectomy with Roux-en-Y reconstruction
    c. Percutaneous placement of a lesser sac drain
    d. Distal pancreatectomy

Answer to Question 3: (d) Distal pancreatectomy. In this scenario with a complete transection of the duct and drain, the pancreas is unlikely to heal without complications of obstruction, pancreatitis, leak, and/or pseudocyst. Non-operative management is likely to lead to these problems. It would be difficult to place a percutaneous drain at the site of injury without damage to the overlying organs and the drain is unlikely to achieve healing of the duct transection. Since the duodenum and head of pancreas are uninjured, the Whipple procedure is inappropriate, and the injury does not warrant the risks of this procedure. The most effective and safest procedure overall is a distal pancreatectomy, with or without a postoperative drain in the bed of resected tail of pancreas.

## References

1. Cunningham RM, Walton MA, Carter PM. The major causes of death in children and adolescents in the United States. N Engl J Med. 2018;379(20):2468–75.
2. Mikrogianakis A. Penetrating abdominal trauma in children. Clin Ped Emerg Med. 2010;11(3):217–24.
3. Moore EE, Cogbill TH, Malangoni M, Jurkovich GJ, Champion HR. Scaling system for organ specific injuries. The American Association for the Surgery of Trauma. http://www.aast.org/Assets/56ef079d-229c-45f2-9b18-c3825e450e65/633867256925730000/injuryscoringtables-pdf. Accessed 20 Oct 2021.
4. Paz MS, Mendez MD. Waddell Triad. StatPearls Publishing LLC, 2021. https://www.ncbi.nlm.nih.gov/books/NBK537094/. Accessed 30 Oct 2021.
5. Lane WG, Dubowitz H, Langenberg P, Dischinger P. Epidemiology of abusive abdominal trauma hospitalizations in United States children. Child Abuse Negl. 2012;36(2):142–8.
6. Trokel M, DiScala C, Terrin NC, Sege RD. Patient and injury characteristics in abusive abdominal injuries. Pediatr Emerg Care. 2006;22(10):700–4.
7. Lindberg D, Makoroff K, Harper N, et al. Utility of hepatic transaminases to recognize abuse in children. Pediatrics. 2009;124(2):509–16.

8. Stylianos S. APSA Trauma Committee. Evidence-based guidelines for resource utilization in children with isolated spleen or liver injury. J Pediatr Surg. 2000;35:164–9.
9. Burch JM, Feliciano DV, Mattox KL. The atriocaval shunt, facts and fiction. Ann Surg. 1988;207(5):555–68.
10. Heo Y, Kim DH. Medial visceral rotations: the Cattell-Braasch vs. the Mattox maneuvers. Trauma Image Proceed. 2020;5(1):39–41. Videos of these maneuvers can be found at http://www.traumaimpro.org/journal/view.php?number=83#supplementary-material. Accessed 30 Oct 2021
11. Anand T, Obaid O, Nelson A, et al. Whole blood hemostatic resuscitation in pediatric trauma: a nationwide propensity-matched analysis. J Trauma Acute Care Surg. 2021;91:573–8.
12. Hamele M, Aden JK, Borgman MA. Tranexamic acid in pediatric combat trauma requiring massive transfusions and mortality. J Trauma Acute Care Surg. 2020;89(2S Suppl 2):S242–5.
13. Branco BB, Naik-Mathuria B, Montero-Baker M, et al. Increasing use of endovascular therapy in pediatric arterial trauma. J Vasc Surg. 2017;66:1175–83.

# Thoracic Trauma

**Aaron Price and Robert Ricca**

**Key Points**

1. Thoracic trauma comprises a small percentage of trauma admissions in the pediatric population, however, when present, accounts for significant increase in morbidity and mortality.
2. It is essential to quickly diagnose thoracic injury in the pediatric trauma patient, as outcomes improve dramatically with expedient and accurate diagnosis.
3. A consistent, stepwise approach to the diagnosis of thoracic trauma can aid in achieving diagnosis while minimizing missed injury.
4. Thoracic trauma can be grouped into two main categories by mechanism, blunt vs penetrating injury.

## Algorithm to Guide Diagnosis and Treatment of Thoracic Trauma in Children

When a pediatric patient presents to the trauma bay with high index of suspicion for thoracic trauma, it is imperative to perform a complete physical examination in a systematic approach so as to not miss an underlying injury. The following is an

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outline for the evaluation of the pediatric trauma patient with respect to presumed thoracic trauma. Children who sustain a thoracic injury can have significant physiologic effects seen in the primary trauma survey.

1) Thoracic trauma can be grouped into two main categories by mechanism, blunt vs penetrating injury, and then further organized by a systematic approach.
    a) Blunt thoracic injury
        i) Skeletal injury
            (1) Rib fracture
            (2) Sternal fracture
            (3) Thoracic spine fracture
        ii) Cardiac/vascular injury
            (1) Myocardial contusion
            (2) Pericardial effusion/tamponade
            (3) Traumatic aortic injury
        iii) Pulmonary/pleural injury
            (1) Pulmonary contusion (most common)
            (2) Pneumothorax/hemothorax
            (3) Diaphragmatic rupture
    b) Penetrating thoracic injury
        i) Pulmonary/pleural injury
            (1) Pneumothorax/hemothorax
            (2) Pulmonary laceration
            (3) Pulmonary contusion
            (4) Diaphragmatic laceration
        ii) Cardiac/vascular injury
        iii) Tracheal/esophageal injury
2) Initial management of thoracic trauma upon arrival of child to trauma bay
    a) Patient evaluated using ATLS protocol.
        i) Patient airway patency established and protected as necessary
        ii) Evaluation of breathing and ventilation
        iii) Circulatory status evaluated
        iv) Patient evaluated for injury requiring emergent, life-saving intervention
    b) After the initial primary survey is complete, the secondary survey begins and can be accomplished in a top-down fashion.
        i) Assess head and neck for blunt vs penetrating injury.
        ii) Evaluate for tracheal positioning with respect to midline.

- iii) Assess for jugular venous distension.
- iv) Watch chest rise and fall for symmetry.
- v) Roll patient to evaluate back for injury.
- vi) Evaluate remainder of patient for further injury (abdomen/pelvis/extremities).

c) After establishment of patient stability, a chest x-ray is the initial imaging modality of choice for workup of thoracic traumatic injury.

- i) Assess for any evidence of acute traumatic osseous injury.
  - (1) Evidence of sternal fracture should prompt 12-lead ECG to assess for myocardial pathology/myocardial contusion.
  - (2) Bedside ultrasound is also useful to assess for pericardial effusion in a setting of suspected blunt trauma to the sternum.
- ii) Evaluate mediastinum for evidence of widening.
  - (1) Widening of mediastinum could indicate traumatic great vessel injury, tracheobronchial injury, or injury to the esophagus.
  - (2) Warrants CT imaging for further evaluation of vasculature.
  - (3) The thymus may be mistaken for a widened mediastinum in young infants.
- iii) Evaluate for symmetry of the airway and pulmonary system.
  - (1) Check for large volume hemothorax/pneumothorax.
    - (a) Due to increased pliability of pediatric chest wall, higher amounts of energy can be transferred to the underlying viscera, and thusly more serious injury can be identified without the presence of obvious rib fracture or sternal fracture.
    - (b) Presence of large volume hemothorax or pneumothorax warrants immediate drainage and decompression as the increased tissue pliability of the pediatric population can lead to significant mediastinal displacement and cardiovascular collapse in rapid fashion.
- iv) Diaphragmatic evaluation.
  - (1) Deepening of costophrenic angle can be indication of non-readily visible pneumothorax on supine chest x-ray.
  - (2) Presence of abdominal content displacement into chest warrants exploration for evaluation of traumatic diaphragmatic rupture.
  - (3) Free air identified sub-diaphragmatically should prompt close attention to evaluation of the abdomen.

## Discussion of Specific Injuries in Respect to Thoracic Trauma

1) Pulmonary contusion
    a) Most common pediatric thoracic injury encountered (53%)
    b) More frequently encountered via blunt trauma
    c) Typically presents with delayed maximum symptomatology
    d) Treatment focused on maintaining adequate gas exchange via pulmonary toilet vs more aggressive ventilatory strategies in severely ill trauma patients
    e) Typically resolves without further complication
2) Pulmonary laceration
    a) Most commonly presents via penetrating mechanism.
    b) Patients can present with hemoptysis, respiratory compromise, and hemodynamic instability.
    c) Will commonly present with associated hemothorax/pneumothorax.
        i) Treatment initially calls for chest tube decompression and drainage of the affected hemithorax.
        ii) Presence of persistent bleeding, significant air leak, or massive hemoptysis warrants surgical exploration and intervention.
3) Hemothorax/pneumothorax
    a) Hemothorax
        i) Typically, as a result of injury to the lung parenchyma or intercostal vascular supply
        ii) Identified initially with chest x-ray during trauma evaluation
    b) Pneumothorax
        i) Commonly caused by disruption of lung parenchyma
        ii) Identification with chest x-ray during trauma evaluation
            (1) The exception to this would be tension pneumothorax, which is a clinical diagnosis.
                (a) Tension pneumothorax is associated with absent breath sounds, decreased oxygenation, and hemodynamic instability.
                (b) Emergent decompression followed by chest tube placement is treatment of choice.
    c) Large volume hemo/pneumothorax or symptomatic hemo/pneumothorax initially treated via chest tube placement for evacuation and decompression
        (1) Chest tube size dependent upon age of patient and presumed hemothorax vs pneumothorax.
        (2) Chest tube placement most commonly occurs in the fifth intercostal space at the anterior axillary line.

            (3) Indications for surgical intervention include chest tube placement with immediate return of >20 cc/kg of output, 2–4 cc/kg/h over a 6-h period, or large volume hemorrhage.
            (4) Current trends in management show increased usage of video-assisted thoracoscopic surgical exploration and intervention to assess the affected hemithorax for appropriate drainage and evaluation of associated injury.

4) Rib fractures

    a) Less common in children due to increased compliance of chest wall
    b) Presence of rib fractures in pediatric trauma typically found to have higher rates of associated injury and higher trauma index scores

        i) First and second rib fractures have higher association with both higher-energy mechanism and cervical spine injury.

    c) Treatment directed at analgesia and aggressive pulmonary toilet

        i) Multimodal pain control including oral and IV medications along with possible use of epidural anesthesia and intercostal nerve blocks to provide adequate analgesia in order to perform pulmonary toileting treatments can be used.
        ii) Typically, no surgical intervention indicated.

5) Thoracic vascular injury

    a) Blunt mechanism is most common with pediatric thoracic aortic injury.

        i) Abnormal distal pulse exam may be seen on physical exam.
        ii) Unexplained hypotension in a setting of blunt thoracic injury should raise suspicion for aortic injury.
        iii) Widening of mediastinum may be seen on chest x-ray along with blunting of the aortic knob, rightward tracheal deviation, or cranial shift of the left mainstem bronchus.
        iv) CTA may be utilized for further evaluation and characterization in the setting of hemodynamic stability.
        v) Hemodynamic instability with evidence for possible aortic injury indicates need for surgical intervention.

            (1) Bedside transthoracic echocardiography can be utilized to assess for pericardial effusion and cardiac tamponade.

                (a) May be temporized by pericardiocentesis while awaiting operative intervention.
                (b) If hemothorax or pneumothorax is evident, chest tube placement may be used for decompression and drainage.

6) Cardiac injury

    a) Myocardial contusion

        i) Most common cardiac injury identified in pediatric population
        ii) Can present with cardiac arrhythmia
        iii) Workup should include ECG, appropriate lab work, and possible echocardiography

    b) Pericardial tamponade

        i) Higher association with penetrating injury vs blunt mechanism.
        ii) Presents with Beck's triad.

            (1) Jugular venous distension/increased venous pressure
            (2) Decreased arterial pressure
            (3) Muffled/decreased heart sounds

        iii) Can be confirmed with bedside FAST exam.
        iv) Definitive treatment involves pericardial window creation for drainage.

            (1) Can be temporized by pericardiocentesis prior to definitive management

7) Mediastinal injury

    a) Tracheobronchial injury

        i) Can be associated with shortness of breath, subcutaneous emphysema, pneumomediastinum, hemoptysis, stridor, or pneumothorax
        ii) Radiographic findings

            (1) Pneumomediastinum
            (2) Pneumothorax
            (3) Elevation of hyoid bone
            (4) Obstructed air-filled bronchus

        iii) If identified, surgical intervention indicated for repair

            (1) Endotracheal intubation should be obtained via bronchoscopic assistance so as to not worsen the injury.

    b) Esophageal injury

        i) Most commonly found with penetrating injury mechanism.
        ii) May be associated with nausea, emesis, vague mid-back pain, shortness of breath.
        iii) Chest x-ray may reveal pneumomediastinum and hydrothorax.
        iv) With suspicion of esophageal injury, contrasted swallow study is imaging modality of choice.

            (1) Endoscopy may be utilized if contrasted swallow study provides inadequate visualization and high suspicion remains.

        v) Treatment calls for operative intervention to control spillage along with antibiotic therapy to prevent infection.

    (1) In critically ill patient, staged procedure to obtain source control followed by definitive operation may be required.

8) Diaphragmatic injury
    a) Left diaphragm more commonly injured than right.
    b) Uncommonly found with blunt mechanism, must be ruled out in penetrating trauma in the thoracoabdominal box.
    c) Chest x-ray may reveal evidence of abdominal viscera within the affected hemithorax.
    d) Typically associated with polytrauma in the pediatric population.
    e) Thorough surgical evaluation and repair warranted at time of discovery.

## Additional Notes

Pediatric thoracic trauma is less common than trauma to other regions of the body, but when it occurs can carry significant morbidity and mortality. In particular, providers must be vigilant to detect signs of life-threatening conditions that require rapid action to correct. These are airway compromise, pneumothorax, hemothorax, pericardial tamponade, and hemorrhagic shock. Following the guidelines of Advance Trauma Life Support from the American College of Surgeons gives the clinician the chance to systematically detect and treat these conditions.

A relatively recent development in the treatment of thoracic trauma with cardiorespiratory failure is emergency extracorporeal membrane oxygenation [1]. For some thoracic aortic injuries, catheter-based methods of treatment may offer an alternative to thoracotomy [2]. Where available, hybrid operating rooms will allow a combination of both catheter-based therapy and traditional operative trauma care. After thoracic trauma, fixation of fractured ribs is a technique being used with increasing frequency in adult patients. This technique is beginning to be used in children as well [3].

## Review Questions

1) A 6-year-old male is brought into the Emergency Department after being reportedly struck by a motor vehicle while playing in an apartment parking lot. Upon arrival the patient is noted to be alert and crying, initial vitals with HR at 135 and blood pressure 110/65. The patient's oxygen saturation is noted to be 94% on room air. On secondary survey, the patient is noted to have abrasions with ecchymoses to the right chest wall with no obvious deformity. The patient is found to have slightly diminished breath sounds on the right compared to left side. Chest x-ray is obtained in the trauma bay revealing a significant right-sided pneumo-

thorax without effusion or mediastinal shift or widening. What is your most appropriate next step in the treatment of this patient?

a. Right-side tube thoracostomy for decompression of the right chest.
b. Chest computed tomography to detect aortic dissection.
c. Transport to the operating room for right-side lateral thoracotomy.
d. Emergency department thoracotomy.

Answer: (a.) Right-side tube thoracostomy for decompression of the right chest. The child has been diagnosed with a pneumothorax which can rapidly become a life-threatening condition if it progresses to a tension pneumothorax. Given the patient's current hemodynamic stability, it would be appropriate to proceed the secondary survey while setting up for an immediate right-sided tube thoracostomy for decompression of the right chest.

2) A 12-year-old male is brought to the emergency department as a level 1 trauma activation after reportedly sustaining a stab wound to the left back. The patient is evaluated using ATLS protocol and is initially found to have hemodynamically stable patent airway. One penetrating wound is found to the left posterior mid back without external signs of significant hemorrhage. A chest x-ray is obtained revealing what appears to be a left-sided hemothorax with slight mediastinal shift. FAST exam reveals no evidence for pericardial effusion or tamponade. Shortly after, the patient begins to become dyspneic and starts to cough up bright red blood. The patient continues to be hemodynamically stable. What is the appropriate next step in the evaluation and management of this patient?

a. Continuous positive airway pressure mask.
b. Chest computed tomography.
c. Fluoroscopy esophagram swallow study.
d. Transport to operating room for surgical exploration of the left chest.

Answer: (d) Transport to operating room for surgical exploration of the left chest. This child has suffered pulmonary laceration with injury to the tracheobronchial system. With new-onset hemoptysis, it would be appropriate to proceed to the operating room for bronchoscopic-assisted endotracheal intubation followed by surgical exploration of the left chest. Hemorrhage from a pulmonary injury can become life-threatening through airway compromise and shock if not addressed.

3) A 7-year-old female is brought into the trauma bay after being involved in a high-speed motor vehicle collision. The patient was a restrained passenger and denies hitting her head or any loss of consciousness. After initial evaluation, it is noted that there is significant tenderness to palpation of the mid sternum with step-off deformity. AP and lateral views of the chest are obtained revealing a posteriorly displaced mid-sternal fracture. The aortic knob is easily visualized on chest x-ray, and the mediastinum is within normal limits. What are the next appropriate steps for continued evaluation and management of this patient?

a. Transport to the operating room for midline sternotomy.
b. Focused abdominal sonography for trauma (FAST) to assess for pericardial effusion.
c. Operative fixation of sternal fracture.
d. Discharge to home, and return to emergency department if any worsening of breathing.

Answer: (b) Focused abdominal sonography for trauma (FAST) to assess for pericardial effusion. Given the mechanism of injury and chest film findings, it would be recommended to obtain a FAST exam to further assess the pericardium to assess for pericardial effusion. Additionally, obtaining a 12-lead ECG to assess for arrhythmia and troponins would be indicated for completeness. If the patient were to experience any indication for hemodynamic instability, repeat chest x-ray followed by possible CT angiography of the chest to assess for blunt aortic injury could be considered given the amount of force needed to result in sternal fracture.

## References

1. Fenton SJ, Hunt MM, Ropski PS, Scaife ER, Russell KW. Use of ECMO support in pediatric patients with severe thoracic trauma. J Pediatr Surg. 2019;54(11):2358–62.
2. Nicholas NW, Shaw DR, Puppala S. Pictorial review on the endovascular management of paediatric aortic injuries. Br J Radiol. 2020;93(1106):20190017.
3. Polycarpou A, Kim BD. Pediatric surgical rib fixation: a collected case series of a rare entity. J Trauma Acute Care Surg. 2021;91(6):947–50. https://bit.ly/ribfixation. Accessed 2 Nov 2021

## *Further Reading*

Beaver BL, Laschinger JC. Pediatric thoracic trauma. Semin Thorac Cardiovasc Surg. 1992;4(3):255–62.

Holl EM, Marek AP, Nygaard RM, Richardson CJ, Hess DJ. Use of chest computed tomography for blunt pediatric chest trauma: does it change clinical course? Pediatr Emerg Care. 2020;36(2):81–6.

Pearson EG, Fitzgerald CA, Santore MT. Pediatric thoracic trauma: current trends. Semin Pediatr Surg. 2017;26(1):36–42.

Peclet MH, Newman KD, Eichelberger MR, Gotschall CS, Garcia VF, Bowman LM. Thoracic trauma in children: an indicator of increased mortality. J Pediatr Surg. 1990;25(9):961–5.

Reynolds SL. Pediatric thoracic trauma: recognition and management. Emerg Med Clin North Am. 2018;36(2):473–83.

Yanchar NL, Woo K, Brennan M, Palmer CS, Zs Ee M, Sweeney B, Crameri J. Chest x-ray as a screening tool for blunt thoracic trauma in children. J Trauma Acute Care Surg. 2013;75(4):613–9.

# Burns

**Elizabeth J. Hughes and Sigrid Blome-Eberwein**

**Key Points**

1. Burn injuries are responsible for nearly 2500 childhood deaths per year.
2. Smoke inhalation is the most important contributing factor.
3. Burn severity is determined by Total Body Surface Area (TBSA) and depth of burn.
4. Lund-Browder chart is used for estimating percent body surface area (BSA) burn; a child's palm (including fingers) is roughly equivalent to 1% BSA. These calculations guide fluid resuscitation measures in the burn patient.
5. Pediatric burn fluid resuscitation differs from that of adults. Children need glucose in their maintenance fluids in addition to resuscitation fluid, and the urine output goal is 1–2 cc/kg/h, as opposed to 0.5–1 cc/kg/h in adults.

## Outline

1. Epidemiology
    a. Mortality from burns is highest in the pediatric and elderly population.

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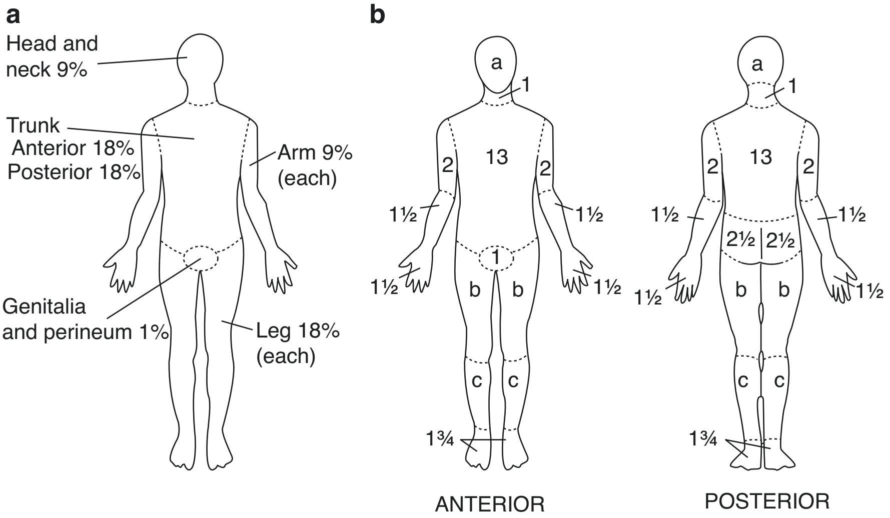
   b. Causes of burns are:
      i. Flame (superheated air)
      ii. Scald (hot liquids), common in children <5 years old
      iii. Contact (hot or cold solid objects), common in toddlers
      iv. Chemical (skin contact with noxious formulas)
      v. Electrical (conduction of electricity through skin and tissues)
   c. Scald and flame burns are most likely type of burn seen in hospital presentations.
      i. These are also the most common type of burn in adolescents, i.e., fireworks, volatile agents.
   d. Providers should have a high index of suspicion for child abuse or neglect in the setting of burn injuries (specific patterns like circumferential extremity with clear demarcation lines, sitz-bath burns, obvious contact identifiers, etc.).
2. Pathophysiology
   a. For purposes of burn classification (see below), layers of skin and soft tissues of the body can be broken down into three groups: epidermis, dermis, and sub-dermis.
      i. Epidermis
         1. *First-degree burn* = red, blanchable skin, painful, no open wound, no scar, i.e., sunburn
      ii. Dermis (corium)) and dermal appendages
         1. *Papillary dermis: superficial partial-thickness second-degree burn* = red, moist/blebs/blisters/bullae, hair follicles intact, painful
         2. *Reticular dermis: deep partial-thickness second-degree burn* = pale/white, blister, hair follicles only partially intact, decreased sensation, heals with scar
      iii. Subcutaneous and subdermal structures
         1. *Third-degree burn* = gray/red/white, dry, leathery/charred, insensate, epidermis sloughs off without blister.
         2. Involvement of structures deep to the subcutaneous fat still constitute third-degree burn. Additional verbage is necessary to accurately describe these types of burns (i.e. muscle, tendon, bone (organ involvement).
         3. These structures may be directly visualized in the wound.
3. Clinical Presentation
   a. Referral criteria from the American Burn Association (ABA)):
      i. Partial-thickness and full-thickness burns >10% TBSA in patients under 10 years old, or greater than 50 years old

   ii. Partial-thickness and full-thickness burns >20% TBSA
   iii. Full-thickness burns >5% TBSA
   iv. Partial-thickness and full-thickness burns involving the face, eyes, ears, hands, feet, genitalia, perineum, or over a major joint
   v. Significant chemical or electrical burns
   vi. Inhalational injury
   vii. Burn injury in patients with pre-existing medical conditions that may complicate treatment and recovery

4. Diagnosis (Fig. 1)

   a. Burn severity is determined by depth of skin injury (degree) and body surface area (% BSA)

      i. In children, the Lund-Browder classification scheme is more accurate than the "Rule of 9's" used for adults and adolescents. The discrepancy between these classification systems is based on a child's relatively large head and small legs when compared to an adult's BSA distribution.


a
Head and neck 9%
Trunk
Anterior 18%
Posterior 18%
Arm 9% (each)
Genitalia and perineum 1%
Leg 18% (each)
b
a
1
13
2
2
1½
1½
1½
1½
1
b
b
c
c
1¾
a
1
13
2
2
1½
1½
2½
2½
1½
1½
b
b
c
c
1¾
ANTERIOR
POSTERIOR

Relative percentage of body surface areas (%BSA) affected by growth

| | 0*yr* | 1*yr* | 5*yr* | 10*yr* | 15*yr* |
|---|---|---|---|---|---|
| a – ½ of head | 9½ | 8½ | 6½ | 5½ | 4½ |
| b – ½ of thigh | 2¾ | 3¾ | 4 | 4¾ | 4½ |
| c – ½ of 1 lower leg | 2½ | 2½ | 2¾ | 3 | 3½ |

**Fig. 1** Estimating percent total body surface area in children affected by bums, (**a**) Rule of "nines" for the rapid estimation of total body surface area burned. In infants, relative surface area of the head is larger, and the tower extremities are smaller. (**b**) Lund-Browder diagram for more precise estimation of burn total body surface area (From US Department of Health and Human Services. Downloaded from https://remm.hhs.gov/burns.htm. On 7 Dec 2013)

    ii. A pediatric patient's palm (including fingers) can be used as a gross estimate of 1% BSA.

  b. Inhalational injury is a potential airway emergency. Consider early intubation with signs of inhalational injury to prevent loss of airway. Signs of *potential* inhalational injury and risk of airway compromise include:

    i. Significant face or neck burns
    ii. Signed eyebrows, nasal hairs
    iii. Carbonaceous sputum
    iv. Hoarseness
    v. Impaired mentation (carbon monoxide poisoning)

5. Initial management

  a. ABCs of trauma

    i. Airway—ensure a definitive airway

      1. Low threshold for endotracheal intubation in setting of inhalational injury, i.e., if the patient inhaled hot gases or significant amounts of smoke, or there are signs of soot involving the oropharynx.
      2. Upper airway is at risk due to thermal injury and chemical injury, and providers should be concerned for progression of airway edema and obstruction.

    ii. Breathing—administer 100% oxygen

      1. Assess for carbon monoxide (CO) poisoning, especially if the patient was involved in a house fire. Pulse oximetry will provide a false reading of oxygen saturation, as CO competes for binding sites on hemoglobin.

    iii. Circulation—it is critical to establish IV access for planned fluid resuscitation, and this should be via non-burned skin if possible.

      1. Two large-bore IVs in upper extremity are preferable.
      2. Central venous access is indicated for severe, large BSA burns, not necessarily before transport to a burn center.

  b. Exposure: it is critical to remove any source of ongoing thermal injury

    i. Remove all clothing and jewelry carefully.
    ii. Brush away dried chemicals.
    iii. Rinse involved sites with copious amounts of tap water, but do not use cold water.
    iv. Cover burns with non-adherent dressings and linens to prevent desiccation and hypothermia. This provides some pain relief (this should occur during transport of patient to tertiary burn center).

c. Fluid, electrolytes, and nutrition

  i. IV access as above, use isotonic crystalloid. Lactated ringer's (LR) is the preferred fluid.

    1. Using colloid fluids, such as albumin, during initial resuscitation poses a risk of respiratory complications related to pulmonary edema.
    2. For children <30 kg, give an additional volume of maintenance fluid *with* glucose, typically D5-1/2 NS. This is due to the danger of severe hypoglycemia early in resuscitation.

  ii. Parkland formula for fluid resuscitation = 4 cc/kg × %BSA 2nd + third-degree burns (first- degree burns do not count for fluid calculation)

    1. Administer half of this volume over the first 8 h. (from the time of injury)
    2. Administer the second half of this volume over the next 16 h.

  iii. A useful endpoint of fluid resuscitation in children is urine output of 1–2 cc/kg/h. Foley insertion for accurate measurement of ins and outs should be considered in children with >15% TBSA burns. This should also be considered in intubated patients, and those with comorbidities complicating the resuscitation of the patient.
  iv. Laboratory examination- CBC, electrolytes, serum glucose, ABG (including carboxyhemoglobin), type and cross-match 10 cc/kg of RBC, CK if very deep burns and/or electrical injury.
  v. Tetanus vaccine administration for prophylaxis in all burn patients.
  vi. Early enteral nutrition is paramount, as moderate and severe burns result in a hypermetabolic state.

    1. Caloric needs in the burn patient: 25 kcal/kg/day plus 30 kcal × %TBSA.

    2. Protein needs: 1 g/kg/day plus 3 g × %TBSA.

    3. Establishment of enteral access (NG, OG, PEG) may be necessary in severe burn patients, and in those who are intubated.

d. Escharotomy is incision of the eschar down to viable subcutaneous fat using electrocautery, usually performed by the burn or trauma specialist, unless delay would jeopardize the extremity or the patient's ventilation. It is indicated for the following: (Fig. 2)

  i. Circumferential deep burns
  ii. Signs of developing compartment syndrome in any extremity: weak pulse, decreased capillary refill, decreased sensation, decreased motor/sensory function, decreased temperature, verify with compartment pressure measurement
  iii. Deep chest/torso burns, especially if there is difficulty achieving adequate ventilation

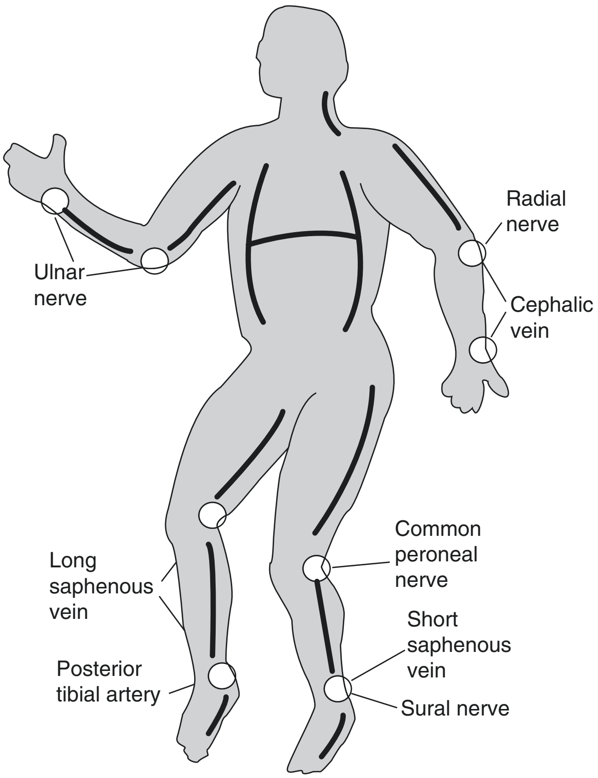

Radial nerve
Ulnar nerve
Cephalic vein
Common peroneal nerve
Long saphenous vein
Short saphenous vein
Posterior tibial artery
Sural nerve

**Fig. 2** Escharotomy lines and anatomic structures that need to be protected when performing escharotomy (Source: Christopher Coppola)

e. Fasciotomy—by surgical specialist, indicated for:
   i. Signs of compartment syndrome that persist after escharotomy
   ii. Burns associated with crush injury, skeletal trauma, or high voltage electrical injury
f. Ruptured bullae should be debrided, under appropriate pain control or sedation at the bedside, as part of the initial assessment.

6. Treatment
   a. The first step in burn assessment and treatment is the cleaning of burns, which is often necessary to perform under sedation.
      i. Administer analgesic, then clean the burn, being sure to remove all loose blisters and necrotic skin.
      ii. Daily, clean burns with warm water and apply fresh dressings, i.e., antimicrobial creams, non-adherent gauze (vaseline gauze, Xeroform, Adaptic, etc.). Cover with soft, absorbent wraps.

b. Topical antibiotics are commonly used (double antibiotic ointment for most partial-thickness burns, 1% silver sulfadiazine for full-thickness burns)
    i. Mupirocin is useful in patients with known MRSA status, though it is very expensive.
    ii. Ophthalmic antibiotic ointment may be used near the eyes.
    iii. Silver sulfadiazine (Silvadene) side effects: neutropenia, thrombocytopenia
        1. Limited penetration of eschar, may inhibit epithelialization
        2. Contraindicated in patients with known sulfa allergy
    iv. Silver nitrate side effects: hyponatremia, hypokalemia, hypochloremia, hypocalcemia
        1. Causes discoloration of skin
        2. Contraindicated in patients with G6PD deficiency—methemoglobinemia causes hemolysis
    v. Mafenide acetate (Sulfamylon) side effects: metabolic acidosis
        1. Application can be very painful
        2. Good penetration of eschar, broad-spectrum coverage for Pseudomonas and GNR proven burn wound infections

c. Systemic antibiotics are not indicated unless there is proven burn wound infection.
d. Homeopathic remedies commonly used in the Plain communities: B&W and burdock leaves.
e. Very deep burns require staged excision and closure:
    i. Initial surgical burn excision and temporary or permanent wound closure should take place <72 h following the burn injury, but only after adequate fluid resuscitation has been achieved, though earlier excision may be practiced with burns <20% TBSA in stable patients.
        1. Human allograft (last up to 4 weeks), Xenograft (porcine, fish; last up to 2 weeks) are used for temporary closure.
        2. Dermal templates are also used for temporary closure in burns with exposed structures and fascia or in very large burn % TBSA.
    ii. Ultimately, autologous epidermal cell coverage is necessary for wound closure: split-thickness or full-thickness skin grafts, cultured epidermal autograft, or cell-spray
        1. Skin grafts incorporate by the principles of inosculation, imbibition, and neovascularization.
        2. Skin grafts may fail to incorporate in areas with poor vascularization, including tendon, bone, and previously irradiated tissue.
        3. Complications of skin grafting: seroma or hematoma formation

a. Prevention: pressure dressing over the graft.

iii. Burn surgery principles:

1. Tangential excision of burned tissue using various instruments, including the Watson blade, Goulian blade, Weck blade and Versajet hydrosurgery, Amalgatome, Dermabrader or ultrasonic debrider.
2. Excision down to viable tissue can be evidenced by punctate bleeding, normal color and texture.
3. To limit morbidity and mortality, each operation should not result in >1 L estimated blood loss (EBL), >20% TBSA burn excision, or >2 h operative time.

7. Unusual Burns
    a. Chemical burns—after any powder is brushed away, large volume of water flush is used to remove all chemical products, as neutralizing agents may cause further injury. The exception to this is with hydrofluoric acid burns, which are neutralized using Calcium gluconate gel repeatedly until the pain subsides.
        i. Acid/alkali
            1. Acid burns cause coagulation necrosis of the soft tissue (battery acid spillage).
            2. Alkali burns result in a deeper penetration of soft tissue due to liquefaction necrosis, resulting in more significant injuries (drain cleaning products).
        ii. Tar burns must be cooled first, then removed using a lipophilic solvent (mineral oil or Vaseline).
    b. Electrical burns, including lightning- the extent of tissue injury is usually underestimated, and the nervous system is the most vulnerable.
        i. Evaluate for rhabdomyolysis and subsequent renal insufficiency with serial CK and urinalysis (UA).
        ii. Treat empirically with sufficient resuscitation, and consider alkalinization of urine.
        iii. Rhabdomyolysis can progress to compartment syndrome.
        iv. Obtain ECG every 8 h and maintain cardiac monitoring for rhythm disturbances.
        v. Multisystem manifestations of electrical injury are possible, including:
            1. MSK—posterior shoulder dislocation, fractures
            2. GI—intestinal perforation, pancreatic or liver necrosis
            3. Neurologic—polyneuritis due to demyelination, quadriplegia, transverse myelitis, cataracts
            4. Cardiopulmonary arrest following ventricular fibrillation

8. Dermatologic Conditions Treated in Burn Centers
    a. Staphylococcal scalded skin syndrome (SSSS), previously known as pemphigus neonatorum, Ritter's disease.
        i. This disorder is mediated by an exotoxin-producing *Staph. aureus* bacterial infection most commonly seen in infants and children.
        ii. Presentation includes diffuse painful erythema with blistering and desquamation, as well as fever, poor intake, and irritability.
    b. Stevens-Johnson syndrome (SJS, 10–30% BSA) and Toxic epidermal necrolysis (TEN, >30% BSA)
        i. These conditions are now viewed as variants of the same disease process.
        ii. The most common etiology is medication adverse reaction (antibiotics, anti-epileptics; top culprits are Penicillin, Bactrim, Dilantin) causing mild to severe erythema, peeling, and blistering of the skin, which can progress to widespread necrosis if not reversed.
        iii. There may be a prodromal period of fever and flu-like symptoms.
        iv. Treatment involves first stopping the suspected medication, being sure to document in the medical record as a severe patient allergy.
9. Outcomes
    a. Complications
        i. Burn wound infections
            1. Seen most commonly in burns with large surface area >30% BSA.
            2. Though more routine use of topical antimicrobial agents has reduced the rate of burn wound bacterial infections, there is no role for use of prophylactic IV antibiotics in treatment of burn patients. Also, the prevalence of Candida infections has increased due to the use of topical antimicrobials.
            3. *Most common organism present in burn wound infection*: Pseudomonas. Other common organisms include Staphylococcus, *E. coli*, Enterobacter. Most common viral organism is Herpes simplex.
            4. *Signs of a burn wound infection*: burn conversion (second degree turns to third degree), green fat, black wound edges, hemorrhage into burn scar, edema.
            5. *Diagnosis*: biopsy is gold standard; culture of burn wound must demonstrate $>10^5$ organisms.
            6. *Treatment*: systemic antibiotics, burn wound excision with *allograft placement.*
        ii. Peripheral neuropathy may be seen as a result of vascular injury and demyelination.
        iii. Hypertrophic scar

            1. These scars are seen more commonly in deep burns with prolonged healing times (weeks), areas of burn that healed by contraction or across flexor surfaces.
            2. Timing: months after initial injury.
            3. Pathophysiology: increased neovascularization, inflammatory response, mechanical stress, fibrocyte invasion.
            4. Treatment involves, topical application of silicone gel, burn scar compression garments, and later injection of steroid directly into the scar.
                a. Novel use of laser treatment for scar treatment and prevention is especially promising in situations where burn scars develop in anatomically challenging areas or there is a lack of surrounding healthy tissue for which to transplant.
            5. Surgical correction of burn scars is a complex reconstructive option involving rotational flap creation along lines of tension, and should not be performed until 1–2 years following the development of hypertrophic scarring and followed by the use of the above measures.
        iv. Heterotopic ossification of tendons requires intensive physical therapy, and may ultimately need surgical intervention, such as tenotomy.
        v. Ulcers
            1. Curling's ulcer: stress-induced: stress-induced gastric or duodenal ulceration seen in severe gastric or duodenal ulceration seen in severe illness, including severe burns, as result of reduced plasma volume. This leads to ischemia and sloughing of the mucosa.
            2. Marjolin's ulcer: squamous cell carcinoma that develops in areas of chronic non-healing burn wounds and scars.
    b. Non-accidental trauma is prevalent in pediatric burn cases and should be carefully screened for at every initial encounter.
        i. Prevalence of child abuse has been documented as high as 15% of pediatric burn injuries.
        ii. Components of the history suspicious for child abuse include conflicting stories, delayed presentation, history of injuries- burns or otherwise.
        iii. Components of the physical exam suspicious for child abuse include sharp demarcation of burn margins, absence of splash marks, involvement of dorsal hands, any scald burn, deep and localized contact burn.

## Narrative Section

The description and management of burns have been documented as early as 3500 years ago, including cave paintings, Egyptian medical books, and treatments developed by Hippocrates. Historically, classification of burn depth and severity has been debated many a time, with just two well-known examples being Guillaume Dupuytren's description of 6 burn classes in 1832, and the more widely used Wilson's description of 3 depths—epidermal, dermal (superficial and deep), and subdermal. Although there have been tremendous strides in the advancement of burn prevention and wound management, thermal injuries still affect approximately one million children in the USA per year. While most pediatric burn injuries are scald burns and only second degree, a significant percentage of those burns can have short- and long-term morbidity and mortality. About 0.1% of these burns lead to systemic streptococcal or staphylococcal infection with sepsis and limb loss.

A large percentage of pediatric burns leave significant scarring, especially if the palms of the hands are involved, leading to long-term contractures and stunted growth if not addressed throughout childhood and adolescence. If grafting is needed for wound closure, contracture scarring is common across all joints and facial burns can lead to restriction in mimics and alter facial features permanently. Jaw burn scars can get reactivated in adolescence with beginning beard growth and lead to pseudofolliculitis barbae. Large surface area pediatric burn survivors suffer lifelong chronic conditions, including heart disease, joint disease, skin diseases, and psychiatric illness. They have an elevated cancer risk even of solid organ cancers and a decreased life expectancy. The traumatic experience of burn treatment with painful dressing changes can alter the psychosocial development, which leads most pediatric burn centers to cover even small second-degree burns with skin substitutes or dressings that minimize traumatic dressing changes. Lastly, pediatric burns have a high preponderance for child abuse with some typical stigmata that should raise suspicion in the initial medical provider: delayed presentation, blaming another sibling for the injury, clear demarcation lines and simultaneous bruising are just a few to be mentioned here. In summary, the diagnosis and treatment of burn injuries in the pediatric population is a complex endeavor, complemented by the continual development of novel approaches and therapies. Physiologically, the body's hypermetabolic state necessitates aggressive fluid resuscitation, early enteral feeding, and the novel use of anti-catabolic therapies. Implementation of early burn excision and grafting in the 1970s has arguably led to the most significant improvement in overall morbidity and mortality in burn patients, including prevention and control of sepsis.

## Study Questions

1. An 8-month-old female presents to the trauma bay with an estimated 15% partial-thickness burn to the L arm and trunk. Her weight is 9 kg. In addition to obtaining a detailed history from the parents and establishing IV access on the contralateral arm, what is the infant's initial 24-h fluid requirement based on the Parkland formula?

   a. 540 cc, given as 270 cc over the first 12 h, 270 cc over the next 12 h
   b. 540 cc, given as 270 cc over the first 16 h, 270 cc over the next 8 h

c. 405 cc, given as 202 cc over the first 8 h, 203 cc over the next 16 h
d. 405 cc, given as 203 cc over the first 16 h, 202 cc over the next 8 h
e. 540 cc, given as 270 cc over the first 8 h, 270 cc over the next 16 h

Answer: (e) 540 cc, given as 270 cc over the first 8 h, 270 cc over the next 16 h. To apply the Parkland formula for fluid resuscitation, being sure to use only areas of 2nd and third-degree burns in the calculation: 4 × 9 kg × 15 = 540 cc total volume to be given in 24 h. Divide this value in half (540/2 = 270 cc), administering the first half over 16 h, and the second half over the following 8 h.

2. A 24-month-old male who weighs 13.5 kg is transported by helicopter to the nearest regional burn center after being involved in a house fire that occurred over 8 h ago. On exam, the child has sustained 20% TBSA partial-thickness burns to bilateral hands and bilateral lower extremities. Tachycardia is present, mucous membranes are dry, skin turgor is poor diffusely, and there is minimal urine output upon Foley catheter insertion. What is the most appropriate goal endpoint of fluid resuscitation in this patient?

   a. 13.5 cc/h urine output
   b. 27 cc/h urine output
   c. 20 cc/kg bolus as needed
   d. 1.5× maintenance fluids

   Answer: (b) 27 cc/h urine output. In pediatric burn patients, the best endpoint of fluid resuscitation is urine output, specifically 1–2 cc/kg/h. Based on this patient's signs and symptoms on presentation, he is likely severely dehydrated. Barring any medical comorbidities, it would be most appropriate to choose an aggressive resuscitation rate. 2 cc/h × 13.5 kg = 27 cc/h.

3. A pediatric burn patient is admitted to the burn unit with 15% TBSA burns, including a 2% area of dark, leathery skin on the left upper extremity. This area spans approximately 60% of the arm circumference and distal pulses and gross motor and sensation are intact. What is the most appropriate initial therapeutic treatment for this patient?

   a. Bedside escharotomy of the left upper extremity
   b. Liberal application of silver sulfadiazine to all areas of burn
   c. Application of mafenide acetate to the area of eschar on the left upper extremity
   d. Proceed to OR for burn excision and temporary grafting

   Answer: (c) Application of mafenide acetate to the area of eschar on the left upper extremity. Mafenide acetate is a topical antibiotic in the form of a cream that has excellent penetration of eschar, while silver sulfadiazine does not. Escharotomy is not indicated at this time, given that the patient is neurovascularly intact, and operative intervention should be delayed to allow for adequate resuscitation.

## Further Reading

Bernabe KQ, Gourlay DM, Gonzalez R, et al. “Burns”. *Pediatric Surgery NaT*, American Pediatric Surgical Association. APSA Webapp. 2020. www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829114/all/Burns.

Herndon D. Total burn care. 5th ed. Philadelphia: Elsevier; 2017.

Hussain A. Surgical treatment of acute burns. Wounds UK. 2013;9(4):1–5.

Jeschke MG, Shahrokhi S, Finnerty CC, et al. Wound coverage technologies in burn care: established techniques. J Burn Care Res. 2018;39(3):313–8.

Klifto KM, Asif M, Hultman CS. Laser management of hypertrophic burn scars: a comprehensive review. Burns Trauma. 2020;8:tkz002. https://doi.org/10.1093/burnst/tkz002.

Townsend CM, Beauchamp RD, Evers BM, et al. Sabiston textbook of surgery. 20th ed. Philadelphia: Elsevier; 2017.

# Cervical Trauma

**Joshua Senter and Aileen P. Wertz**

**Key Points**

1. Providers should maintain high clinical suspicion in the setting of any trauma to the neck in children due to the potential for significant injury to underlying vasculature, nerve structures, aerodigestive tract, or spine with limited symptoms.
2. Management of penetrating cervical trauma is often dictated by which zone of the neck is injured (Zone I, II, or III), with injuries to Zone I and III warranting computed tomography (CT) angiography prior to surgical exploration.
3. Suspected injuries to the aerodigestive tract are best evaluated by combined diagnostic techniques including CT, fluoroscopy, esophagoscopy, and bronchoscopy.
4. Choice of imaging of the cervical spine is complex with plain radiographs, CT, and magnetic resonance imaging (MRI) each conferring unique benefits and pitfalls. MRI remains the gold standard for cervical spine imaging.

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1. Epidemiology:
    a. Eighty to ninety percent of external cervical injuries in children are caused by blunt trauma, whereas the majority in adult patients are due to penetrating trauma.
    b. Cervical spine injury is less common in children.
        i. Cervical spine injuries represent 70–80% of all traumatic spine injuries in children.
        ii. Sixty percent of cervical spine injuries occur from the occiput to level of C2.
    c. A total of 1.9 million trauma cases were reported in National Trauma Database from 2002 to 2006, 633 of which were recorded as laryngeal traumas.
    d. About 0.0036% of the laryngeal trauma events occurred in the pediatric population (69 cases).
    e. The most common cause of esophageal injury in children remains iatrogenic (dilation, placement of endotracheal or nasogastric tubes, or endoscopy).
    f. Esophageal injury secondary to blunt trauma is exceedingly rare.
2. Etiology:
    a. Blunt trauma
        i. Rapid deceleration
        ii. Clothesline injury
        iii. Interpersonal violence
        iv. Strangulation
    b. Penetrating trauma
        i. Gunshot wound
        ii. Knife
        iii. Bites
        iv. Fall on sharp object
3. Pathophysiology:
    a. Given less total body weight in children, less force is applied to head/neck region in cases of rapid acceleration/deceleration.
    b. Increased risk for traumatic brain injury and cervical spinal cord injury is secondary to relatively larger head size and less strength of cervical musculature.
    c. Pediatric larynx sits higher in the neck than the adult larynx.
        i. C3–C5 in pediatric patients versus C6–C7 in adults.
        ii. Mandible offers protection to the pediatric larynx.
    d. Pediatric laryngeal cartilages less ossified, lessening risk of fracture.

   e. Loose adherence of mucosa to laryngeal cartilages creates predisposition to edema, soft tissue damage, and potential hematoma formation.
   f. Given relative narrow caliber of pediatric airway, more susceptible to airway obstruction in presence of edema or hematoma (Poiseuille's law).

4. Emergent Management:
   a. ABCDEs per Advanced Trauma Life Support (ATLS) protocol
      i. Airway and cervical spine protection
      ii. Breathing and ventilation
      iii. Circulation
      iv. Disability
      v. Exposure
   b. Detailed head and neck examination
   c. Consider fiberoptic nasopharyngolaryngoscopy

5. Notable Symptoms and Examination Findings:
   a. Signs *suggestive* of vascular injury
      i. Rapidly expanding or pulsatile hematoma, severe hemorrhage, shock refractory to fluid resuscitation, decreased or absent pulse, vascular bruit/thrill, neurologic deficit
   b. Signs and symptoms *suspicious* for vascular injury
      i. Minor hemoptysis, hematemesis, dysphonia, dysphagia, subcutaneous or mediastinal air, non-expanding hematoma.
   c. Signs and symptoms *suggestive* of laryngeal injury
      i. Respiratory distress, dysphagia, hemoptysis, stridor, impaired phonation, and subcutaneous emphysema.
   d. Signs *suspicious* for laryngeal injury
      i. Neck ecchymosis, inflammation, loss of laryngeal prominence, hyoid elevation.
   e. Signs *suspicious* for esophageal injury
      i. Dysphagia, odynophagia, hemoptysis, neck pain, subcutaneous emphysema, fever, retching.

6. Classification Schemes and Criteria:
   a. Shaefer/Fuhrman Blunt Laryngeal Trauma Classification

      Class 1: Minor endolaryngeal hematoma without detectable fracture.
      Class 2: Edema, hematoma, minor mucosal disruption without exposed cartilage, and nondisplaced fractures.

Class 3: Massive edema, mucosal disruption, exposed cartilage, vocal fold immobility, and displaced fracture.
Class 4: Disruption of anterior larynx, unstable fractures, two or more fracture lines, or massive trauma to laryngeal mucosa.
Class 5: Complete laryngotracheal separation.

b. Zone system of penetrating neck trauma

Zone I: Clavicles to cricoid

i. Innominate vessels, common carotid, subclavian, and vertebral arteries
ii. Brachial plexus
iii. Trachea, esophagus, thoracic duct, and apex of the lung

Zone II: Cricoid cartilage to angle of the mandible

i. Carotid and vertebral arteries, internal jugular vein
ii. Trachea and esophagus
iii. Nerves: Vagus, phrenic, hypoglossal, accessory, ansa cervicalis, great auricular, transverse cervical

Zone III: Angle of mandible to the skull base

i. Carotid and vertebral arteries
ii. Pharynx

c. NEXUS criteria indicating cervical spine imaging should be performed.

i. Midline posterior tenderness, altered level of alertness (including intubation), focal neurologic deficit, and presence of a painful distracting injury.

7. General Radiographic Study Considerations:

i. Plain radiography (XR)

- Less radiation dose than CT

ii. Computed tomography (CT)

- Greater sensitivity than XR
- Required if evidence of atlanto-occipital subluxation on XR

iii. Magnetic resonance imaging

- Gold standard for imaging of the cervical spine
- Allows for evaluation of ligamentous injury
- Time consuming, may require sedation, and patient overall stability may not allow for lengthy imaging

8. Management Based on Specific Injury:

a. Penetrating soft tissue trauma

i. Classically, revolved around anatomic zone(s) of the neck affected (see above description).

ii. Assessment and management of Zone I and III more difficult as access is limited by the clavicles, structures of the thoracic inlet (Zone I), mandible, and skull base (Zone III).

  - CT angiography recommended

iii. Zone II differs in relative ease of access to its structures.

  - In patients with signs or symptoms *suggestive* of vascular injury (see above), surgical exploration warranted.
  - In asymptomatic patients, unlike Zone I and III, management may consist of exploration or directed evaluation and serial examinations at the surgeon's discretion.
  - Angiography indicated for Zone II injuries in stable patients with persistent symptomatology suspicious for vascular or neural injury such as persistent bleeding, neurologic deficit (i.e., Horner's syndrome or hoarseness not explained by laryngeal examination).

b. Cervical spine

i. Cervical spine imaging

  - Damage to cervical spine should be considered in setting of significant neck pain, torticollis, suspicious mechanism of injury, neurological abnormalities.
  - Children > age of 3 may avoid imaging based on the NEXUS criteria (see above).
  - Children < age of 3 may avoid imaging if:
    - Glasgow coma scale (GCS) > 13, neurologically intact, no posterior midline cervical tenderness, lack of unexplained hypotension, no evidence of intoxication, and mechanism of injury does not include motor vehicle accident (MVA), fall >10 feet, or non-accidental trauma.

ii. Surgical management and/or fixation required for the following fractures:

  - Atlanto-occipital dislocation
  - Jefferson fracture
  - Type 2 odontoid fractures
  - Hangman's fractures if significantly displaced
  - Teardrop fracture
  - Unstable cervical compression fractures

iii. Cervical collar may be utilized for management of the following fractures:

  - Atlanto-axial distraction
    - May heal in collar, but often requires surgical treatment.

    - Type 1 and Type 3 odontoid fractures
        - Amount of body involvement may escalate management of Type 3 odontoid fractures to placement of Halo or surgery.
    - Stable cervical compression fractures

c. Esophageal injury

  i. Diagnosis

  - Eighty-eight percent of patients with esophageal perforation will display abnormality on chest XR.
  - CT and/or esophagoscopy indicated if negative chest x-ray (CXR) with persistent suspicion for esophageal perforation.
      - Water-soluble oral contrast should be used to avoid inflammatory reaction if a perforation is present.
  - Most seen as iatrogenic injury in neonates after traumatic naso-/orogastric tube placement or intubation of the esophagus.

  ii. Conservative management

  - More often to result in successful outcome than in adult patients.
  - Ten to fourteen days broad spectrum antibiotics.
  - Nil per os or nothing by mouth (NPO) for at least 7 days.
  - Nutrition via nasogastric tube, gastrostomy, or total parenteral nutrition.
  - Imaging must be repeated prior to initiation of PO feeds.
  - Thoracostomy tube placement for management of associated mediastinal collections, pneumothoraxes, or pleural effusions.
  - In neonate with esophageal perforation with false track, conservative management. Place naso-/orogastric tube past perforation under fluoroscopy.

  iii. Surgical exploration indications

  - Massive spillage, persistent leak, or clinical deterioration

d. Laryngotracheal trauma

  i. Blunt laryngeal trauma may result in bone or cartilage fracture, vocal fold avulsion, arytenoid joint dislocation, soft tissue lacerations, hematomas and/or edema, tracheobronchial rupture, and laryngotracheal separation.
  ii. Flexible nasopharyngolaryngoscopy required to determine extent of injury and appropriate management (patient stability allowing).
  iii. CT neck often indicated to rule out fracture (patient stability allowing).
  iv. Classically, management based on Shaefer/Fuhrman classification (outlined above) of blunt laryngeal trauma, but its utility in children is unclear (see note below).

- Class 1 injury
  - Conservative management
- Class 2, 3, and 4 injuries
  - Definitive airway management (operative laryngoscopy/surgical examination of the airway, possible intubation, possible tracheotomy)
- Class 5 injury
  - Immediate tracheotomy, direct laryngoscopy and esophagoscopy, neck exploration

v. Indications for surgical repair

- Exposure of laryngeal cartilage – chondritis may develop if mucosa not closed over cartilage where possible
- Fractured thyroid and/or cricoid cartilage – repair utilizing plates, screws, and/or sutures
- Vocal fold immobility
- Concern for tracheal rupture or laryngotracheal separation

vi. Conservative management

- Humidified air, steroids, and close observation with serial fiberoptic laryngoscopies.
- Maintain a low threshold for escalation of management to the OR if worsening symptoms or examination.
- If no progression of signs or symptoms, discharge may be considered after 24–72 h observation.

vii. See Ref. [1] for management algorithm of blunt laryngotracheal trauma in pediatric patients.

## Additional Notes

The Schaefer/Fuhrman classification strongly advocates for CT neck across all classes of injury. In children, false-negative CT scans are more frequent, secondary to lack of ossification of laryngeal cartilage. Maintenance of high clinical suspicion despite a negative CT scan is necessary to avoid dismissing cases of clinically significant injury. In the presence of significant symptoms or physical examination findings, operative endoscopy should be performed. Delay to obtain a CT scan or attempt bedside laryngoscopy may be catastrophic in the pediatric population prone to rapid respiratory deterioration.

While historically, any penetrating trauma through the platysma in Zone II required neck exploration, now, a more conservative approach is used based on the clinical findings and imaging as discussed above.

## Study Questions

1) CT angiography is warranted prior to surgical exploration in penetrating trauma to which zone of the neck?
   a. Zone I
   b. Zone II
   c. Zone III
   d. Zone I and Zone III

Answer to Question 1: Choice "d" represents the correct answer. CT angiography is warranted in penetrating trauma to Zone I and III due to relatively difficult surgical exposure and presence of major cervical vessels.

2) Which imaging modality should be utilized if there is concern for ligamentous injury in cervical spine trauma?
   a. MRI cervical spine
   b. CT cervical spine
   c. CT neck w/ angiography
   d. Plain radiograph

Answer to Question 2: Choice "a" is the correct answer. MRI provides superior soft tissue resolution and remains the gold standard for cervical spine imaging, particularly with suspicion of ligamentous injury (Fig. 1).

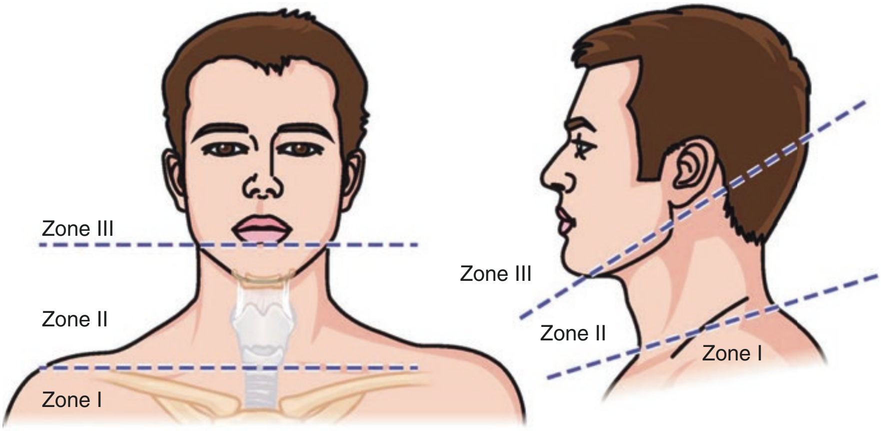

**Fig. 1** Zones of the Neck. (Source: used with permission from Juca Moscardi MF, Pimentel SK Silva FC, Novais JR, Marttos A. Penetrating neck trauma, In: The trauma golden hour, Springer, London, 2020, Downloaded 11 Aug 2020 from https://link.springer.com/chapter/10.1007/978-3-030-26443-7_11)

## Bibliography

1. Cheng J, Cooper M, Tracy E. Clinical considerations for blunt laryngotracheal trauma in children. J Pediatr Surg. 2017 May;52(5):874–80.
2. Alao T, Waseem M. Neck trauma. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021.
3. Bailey BJ, Johnson JT, Newlands SD. Head & neck surgery—otolaryngology. Philadelphia, PA: Lippincott Williams & Wilkins; 2006.
4. Bluestone CD, Healy GB, Simons JP, Cohen MS, Mandell DL, Simons JP. Trauma to the mouth, pharynx, and esophagus in children. In: Bluestone and stool's pediatric otolaryngology. Shelton, CT: People's Medical Publishing House-USA; 2014. p. 1355–62.
5. Bluestone CD, Healy GB, Simons JP, Kelley P. Injuries of the neck. In: Bluestone and stool's pediatric otolaryngology, vol. Vol 2. fifth ed. Shelton, CT: People's Medical Publishing House-USA; 2014. p. 1851–9.
6. Bluestone CD, Healy GB, Simons JP, et al. Diagnosis and management of pediatric laryngotracheal trauma. In: Bluestone and stool's pediatric otolaryngology. Shelton, CT: People's Medical Publishing House-USA; 2014. p. 1627–33.
7. CDC. Web-based injury statistics query and reporting system (WISQARS). Atlanta, GA: US Department of Health and Human Services, CDC; 2009.
8. Copley PC, Tilliridou V, Kirby A, Jones J, Kandasamy J. Management of cervical spine trauma in children. Eur J Trauma Emerg Surg. 2019;45(5):777–89.
9. Ford HR, Gardner MJ, Lynch JM. Laryngotracheal disruption from blunt pediatric neck injuries: impact of early recognition and intervention on outcome. J Pediatr Surg. 1995;30:331–5.
10. Fuhrman GM, Stieg FH III, Buerk CA. Blunt laryngeal trauma: classification and management protocol. J Trauma. 1990;30:87–92.
11. Garton HJ, Hammer MR. Detection of pediatric cervical spine injury. Neurosurgery. 2008;62(3):700–8.
12. Gold SM, Gerber ME, Shott SR, Myer CM. Blunt Laryngotracheal trauma in children. Arch Otolaryngol Head Neck Surg. 1997;123(1):83–7.
13. Mubang RN, Sigmon DF, Stawicki SP. Esophageal trauma. [Updated 2020 Aug 23]. In: StatPearls [Internet]. Treasure Island (FL), StatPearls Publishing; 2021. https://www.ncbi.nlm.nih.gov/books/NBK470161/.
14. Myer CM III, Orobello P, Cotton RT, Bratcher GO. Blunt laryngeal trauma in children. Laryngoscope. 1987;97(9):1043–8.
15. Merritt RM, Bent JP, Porubsky ES. Acute laryngeal trauma in the pediatric patient. Ann Otol Rhinol Laryngol. 1998;107:104–6.
16. Rozzelle CJ, Aarabi B, Dhall SS, Gelb DE, Hurlbert RJ, Ryken TC, et al. Management of pediatric cervical spine and spinal cord injuries. Neurosurgery. 2013;72(Suppl 2):205–26.
17. Schaefer SD. Primary management of laryngotracheal trauma. Ann Otol Rhinol Laryngol. 1982;91:399–402.
18. Viccellio P, Simon H, Pressman BD, Shah MN, Mower WR, Hoffman JR, et al. A prospective multicenter study of cervical spine injury in children. Pediatrics. 2001;108(2):E20.

# Traumatic Thoracolumbar Spinal Injuries in Children

**Gretchen Floan and Romeo Ignacio**

**Key Points**
1) Thoracolumbar spinal trauma is uncommon in pediatric patients when compared to adults, and pediatric patients have particular risk factors and mechanisms that predispose them to spinal trauma.
2) The initial evaluation and workup of a pediatric spinal trauma patient involves a good neurologic examination and imaging with plain films, computed tomography (CT) scans, and/or magnetic resonance imaging (MRI) scans if indicated.
3) Management of thoracolumbar injuries can be operative or non-operative and depends on the stability of the spine, spinal cord compression, underlying neurologic deficits, and potential for long-term healing.
4) There are a variety of fracture patterns that can occur in the spinal column along with neurologic injuries, and it is important to be mindful that some of these injuries can be associated with injuries to other intra-abdominal and thoracic structures.

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1. Epidemiology:
    a. Incidence:
        i. Spinal trauma is an uncommon injury to occur in pediatric patients when compared to adults, and estimates are around 2–5% of all spinal injury cases.
        ii. Spinal fractures are uncommon as well and represent 5% of all pediatric fractures. The incidence in the United States is 1.99 per 100,000 children and most commonly affects African American children followed by Native American, Hispanic, and Asian children. Pediatric spinal trauma is thought to be slightly more common in males.
        iii. The cervical spine is most susceptible to spinal injury in pediatric patients representing 42% of injuries, followed by thoracic (31%) and lumbar (27%).
        iv. The focus of this chapter is thoracolumbar injuries, which are more common in older pediatric patients, typically 8 years old or greater. At this point in their development, the pediatric spine starts to resemble the adult spine more closely and results in a more typical adult-like injury pattern.
    b. Etiology:
        i. In the United States, motor vehicles collisions (MVCs) are the most common cause of spinal column injury (52%), followed by sports (football, rugby, skiing, sledding, All-terrain vehicle (ATV)) accidents, (27%), falls (15%), and child abuse (1–3%). Penetrating injuries (55–60%) and sports injuries (25–30%) had the highest frequency of spinal cord injuries.
        ii. Injuries from MVC can be prevented or reduced, in the majority of cases, by appropriate seat belts and restraints. ATVs in particular have a substantial contribution to thoracolumbar spinal trauma as their usage has rapidly been increasing in children.
2. Pathophysiology:
    a. Anatomy:
        i. <8 years old:
            1. The pediatric spinal vertebra is more mobile and deformable compared to the adult spine. The vertebrae are cartilaginous and incompletely ossified, and the spinal structure is maintained by lax ligaments and underdeveloped spinal muscles. The facet joints are also oriented more horizontally allowing for more mobility. For these reasons, younger children tend to have spinal cord and ligamentous injuries without vertebral fractures.
            2. Young children have a disproportionate head size in relation to their body, creating an innate instability in the cervical spine.

3. The spinal cord in pediatric patients is less protected as the spinal column elasticity allows it to withstand compression from outside forces and transfer this energy to the underlying neural structures.
4. The spinal column is also more mobile than the underlying spinal cord and dura, resulting in a higher incidence of neurologic injury in younger children when compared to adults.

ii. ≥8 years old:

1. Between the ages of 8–10, the spine matures to the point of resembling an adult spine with similar biomechanics. As such, spinal injuries such as vertebral fractures and ligamentous rupture are more common in older children and adolescence.
2. Cervical spinal injury is more common in younger patients for reasons already mentioned, but as the child ages (>8 years old), injuries in the thoracolumbar region are more commonly seen.

3. Clinical features:

a. Presentation:

i. On initial examination, midline tenderness and ecchymosis have an 87% sensitivity and 75% specificity for thoracolumbar fractures.
ii. "Breathlessness" at the time of injury can be used as another marker suggestive of underlying injury despite negative radiographs.
iii. Thoracolumbar spinal fractures can involve multiple levels 11–35% of the time, so the diagnosis of one fracture requires a thorough evaluation of the rest of the spine.

b. Patterns of injury:

i. In pediatric patients, four patterns of injury can describe spinal column trauma. These patterns are based on whether or not there is a fracture, subluxation, or spinal cord injury.
ii. Younger patients typically have

1. ligamentous injury from subluxation or dislocation without a fracture,
2. spinal cord injury without radiographic evidence of fracture based on their spinal mobility and ligament laxity,
3. fractures with subluxation, and
4. fractures without subluxation.

iii. The adolescent population (10–17 years old) more commonly display these latter injury patterns given the development and maturation of their spine.

   c. Associated thoracic and abdominal injuries:
      i. It is important to be aware of other associated intra-abdominal and intrathoracic injuries that can occur in relation to spinal fractures, in particular the association with Chance fractures and injuries to the small bowel, mesentery, and pancreas. This is important to be mindful of as these abdominal injuries can sometimes have a delayed presentation and cause significant morbidity to the patient.
      ii. Thoracic fractures can also be associated with lung contusion or injury, pneumothorax, hemothorax, or aortic injury. These concomitant injuries occur in up to 42% of thoracolumbar spinal injuries.

4. Diagnosis:
   a. History:
      i. In the process of diagnosing a spinal column injury, history, physical examination, and radiographic studies each plays an essential role.
      ii. Mechanism of injury is important along with a history of head trauma or loss of consciousness.
      iii. Screening for midline pain and paresthesias can also be helpful in older children along with a reported history of bowel or bladder incontinence.
      iv. As previously mentioned, reported "breathlessness" may suggest underlying injury.
   b. Examination:
      i. The examination should first start with an evaluation of Glasgow Coma Scale (GCS).
      ii. Next, an attempt should be made to perform a spinal and neurologic examination, including anogenital reflexes if concerned for spinal cord injury. The neurologic examination should be performed serially to evaluate for worsening or improvement in symptoms.
   c. Radiographic studies: no guidelines exist:
      i. X-rays: When it comes to radiographic studies, no clear guidelines exist. Imaging often begins with Anteroposterior (AP) and lateral X-rays of the entire spine as the initial screening modality; however, plain radiographs may fail to detect a number of spinal fractures.
      ii. CT scan: If the patient had a major trauma or there is a high suspicion for spinal trauma, CT scans should be obtained evaluating the entire spine due to risk for multi-level injuries. CT scans assist in diagnosing bone and soft tissue injuries, epidural hematomas, and ligamentous injuries. CT of the head should also be included if the patient is unconscious, deteriorating, or has experienced head trauma. CT scans can also help identify associated injuries in the chest and abdomen but do risk exposing the pediatric population to radiation.

        iii. MRI scan: MRI scans are sometimes obtained in place of the CT scan to spare the patient of radiation exposure. However, they are not as well suited for detecting osseous abnormalities. An MRI can assist with diagnosing injury to the spinal cord as well as neurologic and ligamentous structures. MRIs can also identify operative lesions such as epidural hematoma and traumatic disk herniation. An MRI scan should be obtained when patients present with neurologic deficits on examination.

5. Treatment:
    a. Initial measures:
        i. Initial management starts with securing the Airway, Breathing, Circulation, Disability, and Exposure (ABCDEs) and beginning resuscitation.
        ii. Vital signs should be monitored, and the spine protected using log roll precautions and cervical spine immobilization.
        iii. It is also important to be mindful that patients with cervical or high thoracic spine injuries may display neurogenic shock or develop respiratory failure.
    b. Operative treatment:
        i. Goals of operative intervention:
            1. If a spinal column injury is diagnosed, management depends on the following:
                a. stability of the spine,
                b. spinal cord compression and underlying neurologic deficits, and
                c. potential for long-term healing.
            2. Approximately 7–30% of thoracolumbar spinal fractures require operative intervention with the goals of achieving fracture reduction, stabilization, and spinal cord decompression. Surgeons are becoming more comfortable with thoracolumbar instrumentation in pediatric populations as it has shown promising outcomes regarding injury correction and kyphosis deformities; however, others feel the extent of instrumentation should be minimized given the possible risk for long-term deformities and growth deficits.
        ii. Classifications:
            1. Several injury classification systems exist in order to describe and guide management for spinal column injury.
                a. A three-column system exists dividing the spine into anterior, middle, and posterior columns. Disruption of two or more columns is thought to cause instability to the spine. This classification system is commonly employed but has not been well validated scientifically.

   b. Another classification system, commonly used outside the United States, classifies by fracture type such as burst, compression, flexion–distraction, and flexion–compression. Certain fracture patterns have a higher likelihood of operative management (i.e., burst fractures), and some are better suited for non-operative management (i.e., compression fractures).

2. Thoracolumbar Injury Classification and Severity Score (TLICS) is a reliable and more comprehensive classification method that guides management for patients that may need operative intervention versus conservative management. This classification system includes morphology, disruption of posterior ligamentous complex, and neurologic involvement (Fig. 1).

   a. Each of these components is given a score based on objective findings, and a score >4 leads to a surgical recommendation.
   b. A score equal to 0–3 leads to conservative or non-operative management.
   c. A score equal to 4 is left to surgeon experience and choice of either operative or non-operative intervention.

| Parameter | Points |
|---|---|
| Injury morphology | |
| Compression fracture | 1 |
| Burst fracture | 2 |
| Translational/rotational | 3 |
| Distraction | 4 |
| Posterior ligamentous complex | |
| Intact | 0 |
| Injury suspected/indeterminate | 2 |
| Injured | 3 |
| Neurologic involvement | |
| Intact | 0 |
| Nerve root involvement | 2 |
| Cord/conus medullaris injury | |
| Incomplete | 3 |
| Complete | 2 |
| Cauda equina syndrome | 3 |

**Fig. 1** Thoracolumbar Injury Classification and Severity (TLICS) score. A score >4 leads to surgical recommendation. A score equal to 0–3 leads to conservative or non-operative management. A score equal to 4 is left to surgeon experience and decision. (Source: Adapted from Lee JY, Vaccaro AR, Lim MR, Oner FC, Hulbert RJ, Hedlund R, Fehlings MG, Arnold P, Harrop J, Bono CM, Anderson PA, Anderson DG, Harris MB, Brown AK, Stock GH, Baron EM. Thoracolumbar injury classification and severity score: a new paradigm for the treatment of thoracolumbar spine trauma. J Orthop Sci, 10(6): 671–675, 2005)

  c. Non-operative treatment:
    i. When there is a stable fracture pattern and an absence of neurologic injury, non-operative management can be safely undertaken. Non-operative management consists of external bracing or body casting.
    ii. This method of management can be pursued in some unstable fractures as long as no neurologic deficit exists and there is a high potential for healing with non-operative treatment.

6. Fracture patterns (Fig. 2):
  a. Compression fractures:
    i. Of the fractures in the thoracolumbar region, compression fractures are the most common in patients younger than 18 years old, most often occurring near the thoracolumbar junction and involving multiple spinal levels.
    ii. The increased incidence of this fracture is due in part to the natural wedge shape of the pediatric vertebral body as well as the kyphosis of the pediatric spine.
    iii. These fractures involve low-energy mechanisms such as falls or sports injuries that cause axial loading and spinal flexion causing collapse of the anterior vertebral body.
    iv. Usually, these fractures have a height loss of <30%, but a height loss ≥50% should raise suspicion for a posterior ligamentous injury and prompt MRI.
    v. Often these compression fractures can be managed conservatively with or without bracing and activity modification. Bracing typically consists of thoracolumbosacral orthosis (TLSO) for 6–8 weeks with good results.

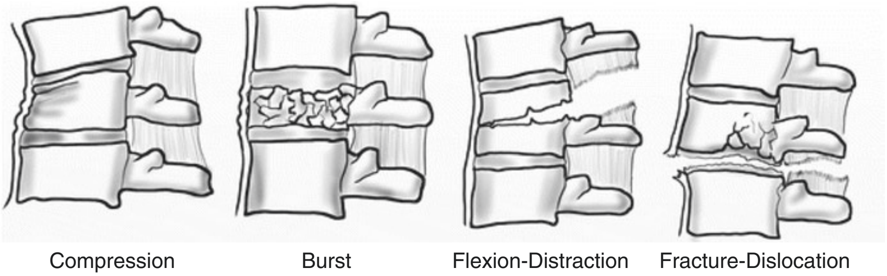

**Fig. 2** Thoracolumbar fracture types in pediatric patients. (Source: Pneumaticos SG, Triantafyllopoulos GK, Lasanianos NG. Thoracolumbar fractures. In: Lasanianos N, Kanakaris N, Giannoudis P, editors. Trauma and orthopaedic classifications. London: Springer, 2015. Used via Copyright Clearance Center RightsLink License No. 52000301)

b. Flexion–distraction fractures (i.e., Chance fractures):

    i. Flexion–distraction fractures, also known as Chance fractures or seat belt injuries, occur with sudden deceleration, and the seat belt migrates toward the spine resulting in direct compression of viscera against spine. The flexion fulcrum is directed onto the anterior vertebral body.
    ii. These fractures most often involve L2 and L3 levels and pose a risk for intra-abdominal injuries (i.e., small bowel, mesentery, and pancreas).
    iii. If the fracture involves the thoracic region, there can also be an injury to thoracic structures involving the lung and aorta (i.e., hemothorax, pneumothorax, lung contusion, and aortic injury).
    iv. Because of the elasticity of the pediatric spine, there is also a risk for spinal cord and dura injury.
    v. These patients may need to be taken emergently to the operating room for concomitant injuries, but an isolated flexion–distraction fracture can be managed non-operatively with TLSO bracing for 8 weeks if the brace provides adequate correction and alignment. Surgical correction may be necessary if the alignment cannot be achieved or maintained. Any ligamentous injury may also require surgical correction.

c. Burst fractures:

    i. Burst fractures are often caused by high-energy mechanisms that cause axial loading and drive the nucleus pulposus into the vertebral body, causing fractures of the anterior and middle columns. With this mechanism, there can be retropulsion of the fragments of the vertebral body involving compression and injury to the spinal cord and dura.
    ii. These fractures are considered unstable and are more often associated with neurologic injury requiring operative intervention for decompression and stabilization.
    iii. A stable burst fracture without neurologic injury can be managed non-operatively with either hyperextension casting or TLSO bracing for 8–12 weeks.

d. Vertebral apophysis fractures:

    i. Children 10–14 years old are most susceptible to apophyseal fractures occurring as a separation of the vertebral apophysis from the vertebral body.
    ii. These fractures most often occur L4 or L5 and can result in posterior herniation of the apophysis into the spinal canal or neural foramen. This injury is comparable to intervertebral disk herniation in adults and produces similar symptoms to include radicular pain after strenuous activity or low-back pain.
    iii. This fracture may not be as readily seen on plain films. CT is a better imaging modality if this fracture is suspected but not identified on plain films. An MRI would also help identify fracture location and extent of herniation.

        iv. The fracture may reduce spontaneously and be managed non-operatively with TLSO bracing for 8 weeks; however, any evidence of neurologic injury needs to be managed surgically.

    e. Slow vehicle crush fractures:

        i. Children under 5 years of age are susceptible to slow vehicle crush injuries that occur when a vehicle is backing out slowly and either pins the child against the vehicle bumper and/or another object causing spinal hyperextension. These injuries can also occur with the child getting run over by a car tire at slow speeds.
        ii. These fractures can have concomitant and potentially major intra-abdominal and thoracic injuries.

    f. Spondylolisthesis/spondylolysis:

        i. Spondylolisthesis/spondylolysis occurs as a traumatic injury, particularly in pediatric athletes that sustain repeated spinal loading and hyperextension. The effect of these biomechanics on an immature spine is fatigue and fracture of the lower lumbar pars. Sports such as gymnastics, weightlifting, and football are contributors to the development of this fracture.
        ii. Treatment can be operative for incapacitating or unresolved pain for >6 months or if there is the presence of neurologic injury or high-grade listhesis.
        iii. Most often, this fracture is managed non-operatively with activity modification.

    g. Spinous/transverse process fractures:

        i. Blunt trauma can result in fractures to spinous or transverse processes. There may be other associated injuries, but a pure injury to these spinal components is managed conservatively with pain control and activity as tolerated.

7. Neurologic injuries:

    a. Patients can also present with neurologic deficits if there is an injury to the underlying neural structures. The American Spinal Injury Association (ASIA) describes spinal cord injuries as complete and incomplete and classifies the level of injury impairment into five categories A–E, A being complete and E being normal.

        i. Complete spinal cord injury, or ASIA grade A, complete lack of all motor and sensory function below the level of the injury.
        ii. Incomplete spinal cord injury retains some motor and/or sensory function below the level of injury.
        iii. The initial spinal cord examination may not represent the true level of the deficit as spinal shock may be present wherein patients lose reflexes below the level of injury. Once spinal shock resolves, a more accurate examination and description of spinal cord injury can be determined.

b. Anterior spinal cord syndrome:
    i. This syndrome results in damage to the anterior two-thirds of the spinal cord involving the corticospinal and spinothalamic tracts but sparing the dorsal columns.
    ii. The mechanism involves hyperflexion and axial loading resulting in injury to the anterior spinal artery and/or injury to the spinal cord from a retropulsed disk or bony fragments.
    iii. Symptoms include loss of pain and temperature sensation along with motor paralysis, sparing proprioception, and vibration sensations.
    iv. This injury has a poor prognosis and potential for functional recovery.

c. Posterior spinal cord syndrome:
    i. Posterior spinal cord syndrome is the opposite of anterior spinal cord syndrome and involves in loss of the posterior one-third of the spinal cord involving the dorsal columns.
    ii. The dorsal columns are responsible for proprioception and vibration sensations which are lost, but motor function as well as pain and temperature sensations remain intact.
    iii. The mechanism involves hyperextension of the spine, particularly the neck and/or direct injury to the posterior spinal cord or disruption of the posterior spinal artery.

d. Central spinal cord syndrome:
    i. Central spinal cord syndrome represents the most common incomplete spinal cord injury, usually resulting from hyperextension.
    ii. The injury involves mainly the central portion of the spinal cord resulting in a motor impartment in the upper and lower extremities. There is a disproportionately greater impact in the upper extremities motor function as the corticospinal tract layers are organized lateral to medial, advancing from the feet to the arms, the arms being more centrally located along the spinal cord. This syndrome can also involve variable degree of sensory loss as well as bladder dysfunction.

e. Brown-Séquard syndrome:
    i. Brown-Séquard syndrome is frequently seen in the cervical spine and is caused by penetrating injury and hemisection of the spinal cord.
    ii. The lateral half of the spinal cord is injured, disrupting the descending corticospinal tracts and ascending dorsal columns. As such, symptoms include ipsilateral loss of motor and proprioception and contralateral loss of pain and temperature sensations.

   f. Cauda equina syndrome:

      i. This syndrome is caused by injury to the cauda equina which is the sac of lumbar, sacral, and coccygeal nerve roots.
      ii. This syndrome may present with several clinical features, including bowel/bladder dysfunction, sensory loss and weakness in lower extremities, saddle anesthesia, and decreased lower extremity reflexes and rectal tone.
      iii. This syndrome is an emergency and requires emergent surgical intervention once diagnosed.

   g. Spinal cord injury without radiographic abnormality (i.e., SCIWORA):

      i. SCIWORA occurs in 6–19% of pediatric patients and is associated with high-speed MVCs, flexion–distraction injures, and slow vehicle crush injuries.
      ii. Incidence is highest among children <8 years old who tend to have a poor prognosis.
      iii. Predisposing factors include heavy head, weak musculature, and flexible spinal column.
      iv. SCIWORA commonly affects the cervical spine, but the upper thoracic spine can also be affected if the watershed vascular supply is disrupted. Despite there being no fracture, SCIWORA is considered an unstable injury due to concern for ligamentous disruption.
      v. With this injury, there is also the potential to develop delayed neurologic deterioration 30 min–4 days after the injury as well as recurrent SCIWORA. These risks are reduced with timely bracing for 12 weeks and spinal precautions.

8. Outcomes:

   a. After a traumatic thoracolumbar spinal injury, the biggest concern for long-term outcomes is scoliotic or kyphotic spinal deformities. These deformities can progress as a result of injury to the growth plate or before an adolescent growth spurt. If the growth plate is not involved in the fracture, however, the probability of developing spinal deformities is unclear. The biggest factor in determining long-term outcomes is the presence of a neurologic deficit at the time of injury. Incomplete neurologic injuries have better outcomes than complete injuries. After a complete neurologic injury, 25% have a full recovery and 60% have partial recovery.
   b. Given the risk of progressive deformity, patients who suffered thoracolumbar spinal injuries should be followed through skeletal development to monitor for deformity development. Those who suffered a spinal cord injury may also develop late syringomyelia and should also be monitored for progression of neurologic symptoms. These deformities can be controlled with bracing, but any progressive deformity or neurologic deficit is an indication for surgical intervention.

9. Prevention:
    a. Although there exist effective management strategies for pediatric spinal trauma, implementing prevention measures should not be forgotten so as to limit these injuries altogether. Motor vehicle collisions are the most common cause of spinal column injury in pediatric patients and as such contribute a significant amount to the injury burden.
        i. Prevention for these injuries includes appropriate restraints and seat belts for children. According to the American Academy of Pediatrics, National Highway Traffic Safety Administration, and Centers for Disease Control and Prevention recommend the following:
            1. Children <2–3 years of age should ride in a rear-facing car seat.
            2. Children >2–4 years of age should transition to a forward-facing car seat with a harness.
            3. Children 4–12 years of age should transition to a booster seat when they exceed the height or weight limit of the car seat manufacturer.
            4. Children may use lap-and-shoulder seat belts when they fit properly and continue to use them at all times for optimal protection.
            5. Children appropriate for lap-and-shoulder seat belts have typically reached 4 feet, 9 inches in height, and are between 8–12 years of age. There is some overlap in ages that are appropriate for the various car seats, so following the manufacturer's guidelines is recommended.
            6. Car and booster seats should never be placed in the front seat as front-seat airbags can injure or kill young children.
            7. The back seat is also safer, and children should sit there at least until they are 12 years of age.
            8. When compared to children using lap-and-shoulder, the practice of these car seat guidelines reduces the risk of injury by 70–80% and reduces the risk of death by 28%. Injuries can be prevented with booster seats by 45% in children 4–8 years old.
        ii. The second most common cause of spinal column injury in pediatric patients is sports. The American Academy of Pediatrics provides recommendations to prevent sports-related injuries of which include the following:
            1. Wearing the appropriate gear for the sports such as helmets and protective padding.
            2. Strengthening muscles and increasing flexibility.
            3. Using proper technique.
            4. Playing safe with rules against headfirst sliding in baseball and softball, spearing in football, and checking in hockey.
            5. Taking time off of at least 1 day per week to allow the body to recover.

iii. ATVs contribute to spinal column injuries as children and adolescents are self-taught and self-operators without adequate knowledge, judgment, or experience.

    1. Prevention measures for ATV use include age restrictions, rider-safety training, and helmet enforcement.
    2. States have varying safety regulations for ATV riders, but there still has yet to be an observed reduction in ATV-related injuries and deaths.

iv. Falls are the third most common cause of spinal column injuries, and they can be prevented with house-proofing safety modifications.

    1. Mayo Clinic offers several effective ways to prevent falls at home, including window locks, preventing windows from opening greater than 6 inches, opening double-hung windows only from the top, and keeping furniture away from windows.
    2. Other safety measures include safety gates at the top of stairs, not allowing children to play unattended on a balcony, installing safety rails on beds, using nightlights, and using nonslip mats in the bathtub.
    3. While away from home, families should seek out playgrounds with shock-absorbing surfaces, only use designated seats in shopping carts, and use wide-based strollers and harnesses to limit injuries in strollers. These safety measures require proactive planning but can be effective in preventing serious injuries and even death in pediatric patients.

## Study Questions

1) A 15-year-old 50 kg female was a restrained passenger in a high-speed motor vehicle collision that hit the median. On examination, she has a seat belt sign and complains of lumbar spinal pain. She has no neurologic deficits. CT scans of the chest, abdomen, and pelvis are obtained with full imaging of the spine. What is the most likely spinal fracture to be found, and what other injury should be evaluated for?

    a. Vertebral apophysis fracture; pancreatic contusion
    b. Burst fracture, spinal cord injury
    c. Flexion–distraction fracture; small bowel ± mesentery injury
    d. Compression fracture; pelvic fracture

Answer to Question 1:

(c) Flexion–distraction fracture; small bowel ± mesentery injury. These types of fractures (i.e., Chance fractures or seat belt injuries) classically occur as a result of MVC and sudden deceleration mechanisms. The anterior vertebral body of L2 or L3, most commonly, acts as a fulcrum around the seat belt which compresses the intra-abdominal organs (i.e., small bowel, mesentery, pancreas) against the spine. It is imperative to rule-out intra-abdominal injury as this can have significant effects on patient morbidity and mortality.

2) A 13-year-old male presents after falling from a 20-foot balcony landing on his sacrum. He progressively developed worsening lumbar back pain and was brought to the hospital for evaluation. On CT scan, he was found to have a burst fracture of the L2 vertebra with protrusion of fractures into the spinal canal. He has decreased sensation and weakness to flexion and extension of bilateral lower extremities. There is a concern for ligamentous disruption based on a CT scan. What Thoracolumbar Injury Classification and Severity (TLICS) score is this patient, and what is the recommended treatment plan?

   a. 5; conservative management with TLSO bracing and activity modification
   b. 6; conservative management with TLSO bracing and activity modification
   c. 7; urgent/emergent operative intervention for spinal cord decompression and spinal column stabilization
   d. 8; urgent/emergent operative intervention for spinal cord decompression and spinal column stabilization

Answer to Question 2:

(c) 7; urgent/emergent operatively intervention for spinal cord decompression and spinal column stabilization. The TLICS scoring system includes morphology, disruption of posterior ligamentous complex, and neurologic involvement. Any score >4 is associated with operative recommendations. This patient's fracture pattern is unstable, not to mention he is showing signs of anterior spinal cord syndrome. For these reasons, the patient should go to the operating room for spinal cord decompression and spinal column stabilization.

3) A 3-year-old female was riding an ATV with her father when they hit unexpected terrain and rolled over. The child was brought to the nearest hospital where plain films and CT scans of the spine were all negative, but despite this, she still has focal pain in the cervical spine. An MRI is obtained showing ligamentous disruption of the cervical spine. What other underlying injury may be present, and what is the most appropriate treatment?

   a. Spinal cord injury without radiographic abnormality (i.e., SCIWORA); operative spinal stabilization
   b. Spinal cord injury without radiographic abnormality (i.e., SCIWORA); bracing and spinal precautions
   c. Missed vertebral fracture; repeat imaging
   d. Damage to trachea; bronchoscopy

Answer to Question 3:

(b) Spinal cord injury without radiographic abnormality (i.e., SCIWORA); bracing and spinal precautions. SCIWORA injuries are most commonly seen in pediatric patients <8 years old and result secondarily from the flexibility of the spine. These injuries present as a spinal cord injury without a radiographic fracture. The cervical spine is the most commonly affected area, and treatment is 12 weeks of spinal bracing and precautions.

## Further Reading

Alexiades NG, Parisi F, Anderson RCE. Pediatric spine trauma: a brief review. Neurosurgery. 2020;87(1):E1–9.

Car seats and booster seats. NHTSA. Retrieved November 27, 2021, from https://www.nhtsa.gov/equipment/car-seats-and-booster-seats

Daniels AH, Sobel AD, Eberson CP. Pediatric thoracolumbar spine trauma. J Am Acad Orthop Surg. 2013;21(12):707–16.

Durbin DR, Hoffman BD, Council on Injury, Violence, and Poison Prevention. Child passenger safety. Pediatrics. 2018;142(5):e20182460. Epub 2018 Aug 30

Follow these tips to prevent your child from falling. Mayo Clinic (2020). https://www.mayoclinic.org/healthy-lifestyle/infant-and-toddler-health/in-depth/child-safety/art-20046124. Accessed 27 Nov 2021.

Goodwin CR, Recinos PF, Jallo GI. Pediatric spinal trauma. Neurosurg Q. 2012;22(2):73–80.

Habermehl N, Minich NM, Malay S, Mahran A, Kim G. Pediatric thoracolumbar spinal injuries in United States trauma centers. Pediatr Emerg Care. 2021.

Huisman TA, Wagner MW, Bosemani T, Tekes A, Poretti A. Pediatric spinal trauma. J Neuroimaging. 2015;25(3):337–53.

Keep child passengers safe. CDC, September 15, 2021. https://www.cdc.gov/injury/features/child-passenger-safety/index.html. Accessed 27 Nov 2021

Lee JY, Vaccaro AR, Lim MR, Oner FC, Hulbert RJ, Hedlund R, Fehlings MG, Arnold P, Harrop J, Bono CM, Anderson PA, Anderson DG, Harris MB, Brown AK, Stock GH, Baron EM. Thoracolumbar injury classification and severity score: a new paradigm for the treatment of thoracolumbar spine trauma. J Orthop Sci. 2005;10(6):671–5.

Mandadi AR, Koutsogiannis P, Waseem M. Pediatric spine trauma. 2021 Aug 13. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021.

McBride AS, Cline DM, Neiberg RH, Westmoreland KD. Pediatric all-terrain vehicle injuries: does legislation make a dent? Pediatr Emerg Care. 2011 Feb;27(2):97–101.

Pediatric thoracolumbar spinal injury: frequently hidden, but not harmless. Relias Media—Continuing Medical Education Publishing. https://www.reliasmedia.com/articles/22734-pediatric-thoracolumbar-spinal-injury-frequently-hidden-but-not-harmless. Accessed 27 Nov 2021.

Sports injury prevention tips. American Academy of Pediatrics, March 20, 2017. https://www.healthychildren.org/English/health-issues/injuries-emergencies/sports-injuries/Pages/Sports-Injuries-Treatment.aspx?_ga=2.267635150.1509095698.1638060574-1140910904.1638060574&_gl=1*ydszvy*_ga*MTE0MDkxMDkwNC4xNjM4MDYwNTc0*_ga_FD9D3XZVQQ*MTYzODA2MDU3My4xLjEuMTYzODA2MDU5MS4w. Accessed 27 Nov 2021.

Srinivasan V, Jea A. Pediatric thoracolumbar spine trauma. Neurosurg Clin N Am. 2017;28(1):103–14. https://doi.org/10.1016/j.nec.2016.07.003. Epub 2016 Nov 1

The TLICS with its subcategories and scoring | download table. https://researchgate.net/figure/The-TLICS-with-Its-Subcategories-and-Scoring_tbl1_258512137. https://www.researchgate.net/figure/The-TLICS-with-Its-Subcategories-and-Scoring_tbl1_258512137. Accessed 27 Nov 2021.

# Pelvic Fractures

**Casssandra Ricketts and Meagan Fernandez**

**Key Points**

(a) Pelvic ring and acetabular fractures are associated with high-energy trauma.

   (i) Mechanism: motor vehicle collisions, pedestrian versus motor vehicle, and falls from height.
   (ii) Association with CNS and abdominal injuries.

(b) Apophyseal avulsion fractures are the result of low-energy trauma.

   (i) Often during sports, i.e., jumping, sprinting.
   (ii) Mostly seen in adolescent patients.

(c) Fracture patterns vary based on status of triradiate cartilage and skeletal maturity.

   (i) Pelvic bones become stronger than ligaments after skeletal maturity.
   (ii) Pediatric patients can have unique fracture patterns.
   (iii) Fractures in adolescent patients often resemble adult-type injuries.

(d) Lower overall mortality in children compared to adult pelvic fractures.

   (i) Pelvic hemorrhage and vascular injuries are less common in children.

      1. May be due to smaller vessel diameter, increased vasoconstriction.

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(e) Acetabular fractures account for <10–15% of all pediatric pelvic fractures.
    (i) Less common than in adults.
    (ii) Can have combined pelvic and acetabular fractures.

1. Clinical anatomy
    (a) Pelvic bones, Fig. 1
        (i) Pubis: superior and inferior rami
        (ii) Ischium: body and tuberosity
        (iii) Ilium: body and wing
    (b) Ligaments
        (i) Pubic symphysis
        (ii) Sacroiliac (SI)
            1. Posterior SI ligaments are the strongest: important factor for vertical stability of pelvis.
        (iii) Iliolumbar: avulsion fracture can be a sign of unstable ring injury.
            1. Spinal sided avulsion: from the L4 or L5 transverse process
            2. Pelvic sided avulsion: from the iliac crest apophysis
    (c) Triradiate cartilage
        (i) Y-shaped growth plate at the junction of three growth centers.
            1. Ischium—ossification center appears at 9 weeks
            2. Ilium—ossification center appears at 16 weeks
            3. Pubis—ossification center appears at 20 weeks

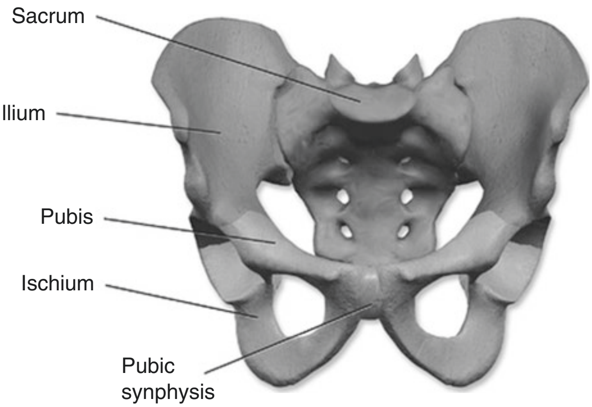

**Fig. 1** The pelvis consists of the pubis, the ischium, and the ilium (Adapted from Bruce Blausen, Blausen Medical Communications, under Creative Commons Attribution 3.0 Unreported license as published on http://en.wikipedia.org/wiki/File:Blausen_0723_Pelvis.png. Downloaded 28 Dec 2013)

      - (ii) Closes around age 14 in boys, 12 in girls.
      - (iii) Adolescent patients with closed triradiate cartilage are at greater risk for pelvic ring injuries.
        1. These fractures behave similar to adult fractures and are treated as such.

2. History and physical examination
   - (a) History
     - (i) Mechanism is either high or low energy.
       1. High energy
          - (a) Motor vehicle accident
          - (b) Pedestrian vs. motor vehicle
          - (c) Falls from height
       2. Low energy
          - (a) Avulsion fractures occur due to sudden traction on a tendon during sports, i.e., sprinting and jumping.
     - (ii) Presenting symptoms
       1. Pelvic pain
       2. Inability to bear weight
   - (b) Physical examination
     - (i) Primary and secondary surveys
     - (ii) Advanced Trauma Life Support (ATLS) evaluation for life-threatening injuries
   - (c) Inspection
     - (i) Inspect the entire body. Logroll to examine the spine and posterior legs.
     - (ii) Assess for Morel-Lavellee lesion: degloving of subcutaneous fat and skin over the fascia.
     - (iii) Assess for rectal/genitourinary open wounds and/or bleeding.
       1. May indicate an open pelvic fracture: rare, but may require diverting colostomy.
   - (d) Palpation
     - (i) Anterior Superior Iliac Spine (ASIS), iliac crests, SI joints, pubic symphysis, and entire spine
     - (ii) Posteriorly directed pressure at the iliac crests/compression of the pelvis
       1. Assess for pain
       2. Assess for stability: excessive mobility may indicate unstable pelvis
   - (e) Complete neurovascular examination of the upper and lower extremities

- (f) Look for associated orthopedic injuries: fractures of long bones, proximal femur, and hip dislocations can occur with pelvic and acetabular trauma
- (g) Thorough urogenital examination
    - (i) Rectal examination required.
    - (ii) Retrograde urethrogram required prior to bladder catheter placement if urethral injury is suspected.

3. Diagnostic imaging
    - (a) Radiographs
        - (i) AP pelvis is part of initial trauma evaluation.
        - (ii) Judet views
            1. 45° internal and external oblique view of the affected hip.
            2. Obtained for acetabular fractures.
        - (iii) Pelvic inlet and outlet views
            1. X-ray beam angled approximately at 40° caudad for the inlet view and cephalad for the outlet view.
            2. Obtained for pelvic fractures.
        - (iv) Radiographs will miss about 50% of all pediatric pelvis fractures.
            1. Computed tomography (CT) scan required in high-energy trauma with positive examination findings.
    - (b) Computed tomography (CT)
        - (i) Improved osseous detail
        - (ii) Better delineation of the fracture pattern
            1. Can also use CT three-dimensional reconstruction, if available at your institution.
        - (iii) Indications for CT
            1. Negative X-rays with concerning history and/or physical examination
            2. Tenderness over the SI joints
            3. Preoperative planning
            4. Spine and pelvic fractures
    - (c) MRI/bone scan
        - (i) Limited value in the acute setting.
        - (ii) Can be useful to diagnose occult fractures or avulsion injuries.
4. Classification of pelvic and acetabular fractures
    - (a) Many classification systems exist.

(b) Limited clinical utility—variability exists in the ability for classification systems to determine treatment and prognosis.
(c) Pediatric pelvic fracture classification.
    (i) Torode and Zieg
        1. Type I: Avulsion
        2. Type II: Iliac wing
        3. Type III: Simple ring fracture with no segmental instability
        4. Type IV: Ring disruption with instability
(d) Adolescent pelvic fractures often resemble adult patterns.
    (i) Tile classification
        1. A: Stable
        2. B: Rotationally unstable or vertically stable
        3. C: Rotationally and vertically unstable
(e) Acetabular fracture classification
    (i) Salter-Harris classification is used with open triradiate cartilage.
    (ii) Salter-Harris classification
        1. Type I: Fracture through physis
        2. Type II: Fracture through physis and metaphysis
        3. Type III: Fracture through physis and epiphysis
        4. Type IV: Fracture through physis, metaphysis, and epiphysis
        5. Type V: Crush injury to physis
    (iii) Fractures in adolescents and skeletally mature patients resemble adult patterns.
        1. Letournel classification
            (a) Elementary types
                (i) Posterior wall
                (ii) Posterior column
                (iii) Anterior wall
                (iv) Anterior column
                (v) Transverse
            (b) Associated types
                (i) Both columns
                (ii) Transverse and posterior wall
                (iii) T-Type
                (iv) Anterior column and posterior hemitransverse
                (v) Posterior wall and posterior column

5. Initial treatment principles
    - (a) Most pediatric pelvic and acetabular fractures can be managed without surgery.
    - (b) Life-threatening injuries need to be managed first: address hemodynamic instability.
    - (c) Treatment often dictated by age.
        - (i) Fractures in skeletally mature patients resemble adult fractures and may be managed like an adult.
        - (ii) Skeletally mature patients have less elasticity in their bone, cartilage, pubic symphysis, and sacroiliac joints.
    - (d) Obtain orthopedic surgical consult.
6. Management of hemorrhage from pelvic fractures
    - (a) Hemorrhage is less common in children than in adults.
        - (i) Smaller vessels with improved ability to vasoconstrict.
        - (ii) Higher rate of simple, stable fractures due to the elasticity and ability of the bones, cartilage, and joints to absorb energy.
    - (b) Associated with Torode and Zieg IV fractures (unstable ring disruptions): iliac vessels often involved.
    - (c) When IV fluid boluses and blood transfusions are required, need to rule out other sources of bleeding, such as abdomen, pelvis, extremity, thorax, and retroperitoneal.
    - (d) Pelvic volume can be initially decreased with pelvic binder or sheet.
        - (i) Sheet wrapped around greater trochanters.
        - (ii) Commercially available pelvic binders should be centered at the level of the greater trochanters.
    - (e) Persistent hemodynamic instability
        - (i) Emergent external fixation
        - (ii) Pelvic angiography: indicated in ongoing hemodynamic instability
            1. Embolization if evidence of pelvic hemorrhage
        - (iii) The protocol for treatment will vary by institution and resources available.
7. Nonoperative treatment
    - (a) Ensure hemodynamic stability
    - (b) Broad indications for nonoperative treatment
        - (i) Avulsion and iliac wing fractures with <2 cm of displacement and stable ring fractures
        - (ii) Most stable acetabular fractures with <2 mm of joint displacement

- (c) Protected weight-bearing initially, followed by progression of weight bearing and physical therapy
    - (i) Indications
        1. Type I: avulsion fractures with <2 cm of displacement
        2. Type II: iliac wing fractures with <2 cm of displacement
        3. Type III: pelvic ring fractures without instability
        4. Closed, stable, congruent acetabular fractures with no intra-articular fragments and <2 mm of displacement
- (d) Bedrest
    - (i) Indications
        1. Type IV pelvic ring fractures with instability and <2 cm of ring displacement
- (e) Skeletal traction
    - (i) Often indicated initially for vertically unstable pelvic fractures.
    - (ii) May also be indicated in unstable, displaced acetabular fractures.
    - (iii) Buck's traction (skin traction) is a short-term, initial alternative.

8. Operative treatment
    - (a) General principles
        - (i) Top priority is hemodynamic stability. This will dictate timing of OR and surgery performed.
            1. External fixation indicated in the hemodynamically unstable patient.
            2. Open reduction and internal fixation (ORIF) is indicated in the hemodynamically stable patient.
        - (ii) Leg length discrepancy <2 cm can be well tolerated in the pediatric patient.
        - (iii) Broad indications for operative treatment
            1. Pelvic fractures with >2–3 cm of displacement and unstable pelvic fractures
            2. Acetabular fractures with >2 mm of displacement
    - (b) External fixation
        - (i) Indications
            1. Hemodynamic instability
            2. Increased pelvic volume
    - (c) Open reduction and internal fixation (ORIF)
        - (i) Pelvic ring indications

            1. Type I avulsion fracture with >2–3 cm of displacement
            2. Type II iliac wing fractures with >2–3 cm of displacement
            3. Type IV pelvic ring fractures with instability

        (ii) Acetabulum indications

            1. Open fractures
            2. Joint displacement >2 mm
            3. Joint incongruity
            4. Intra-articular fragments
            5. Joint instability
            6. Irreducible fracture dislocation

    (d) Spica casting

        (i) May be indicated instead of nonoperative treatment for younger, noncompliant patients.

9. Complications of pelvic and acetabular fractures

    (a) Death

        (i) Rare, often associated with head or organ injury

    (b) Growth disturbance and deformity

        (i) Premature physeal closure

            1. Children <10 years old are at increased risk of more severe growth abnormality.
            2. Can result in acetabular dysplasia.

        (ii) Leg length discrepancy

            1. Increased risk with vertically unstable fractures and displacement >2 cm.
            2. Limb length discrepancy <2 cm is well tolerated and treated nonoperatively.

    (c) Malunion/nonunion

        (i) Rare

    (d) Avascular necrosis of femoral head

        (i) Seen in hip dislocations, acetabular fractures, and proximal femur fractures

    (e) Peripheral nerve injury
    (f) Myositis ossificans, also known as heterotopic ossification (HO)
    (g) Post-traumatic arthritis of the hip

        (i) Increased risk in displaced acetabular fractures

## Discussion

There are significant differences between pediatric and adult pelvic and acetabular injuries. Generally, the prognosis for children with these injuries is better than adults. There is a higher incidence of simple fractures and lower incidence of hemorrhage and death in children. The mechanism is typically motor vehicle accidents or pedestrian vs. auto accidents, which may involve a laterally directed force to the pelvis. This is called a lateral compression injury. Lateral compression injuries tend to be more benign compared to open-book anterior–posterior compression injuries, which are associated with vascular injuries. The pediatric pelvis tends to resist fractures and displacement for various anatomic reasons, similar to long bones. Pediatric bones and joints, such as the SI joint and pubic symphysis, are elastic, and able to resist deforming forces. Pediatric cartilage is able to absorb energy. Additionally, the thick periosteum in children helps maintain the alignment of bones. Due to each of these characteristics, pediatric pelvic fractures are inherently more stable and less severe than their adult counterparts. Importantly, in the setting of high-energy trauma and displaced fractures, children are less likely to hemorrhage and die. This is thought to be due to smaller vessels which can vasoconstrict and prevent massive hemorrhage. Finally, skeletally immature patients have the ability to remodel, making malunion better tolerated than in adults. While pediatric patients do overall better than adults, unstable fractures, especially vertically unstable, must undergo appropriate reduction and stabilization, or patients will have long-term functional deficits.

Acetabular fractures have unique anatomic properties that present challenges. The triradiate cartilage can be injured which leads to growth arrest (premature physeal closure) and subsequent deformity. Younger children are at an increased risk of injury to the growth plate and subsequent acetabular deformity/dysplasia. These deformities are difficult to correct and may require osteotomies to correct the acetabular dysplasia in children over the age of 10.

The role of the pediatric surgeon is to understand the initial management of these injuries, including the history and physical examination, primary and secondary surveys, appropriate imaging, and be able to manage the hemodynamic instability seen in high-energy and unstable fractures. Obtaining a good history and identifying any risk factors for increased hemorrhage is crucial. Physical examination will play a critical role in identifying associated injuries and open fractures. Open pelvic fractures will involve a multidisciplinary team including the pediatric surgery, orthopedic surgery, and possibly urology and gynecology depending on the injury. Appropriate imaging should be ordered, most often a CT scan, as up to 50% of pelvic fractures can be missed on initial radiographs. Once pelvic and/or acetabular fractures are identified, the orthopedic surgery team should be consulted. For persistent hemodynamic instability, emergent external fixation and/or pelvic angiography/embolization is often required. The pediatric surgeon must be vigilant in communicating with a multidisciplinary team in order to optimally manage these patients with hemodynamic instability.

In summary, the prognosis of most pediatric pelvic and acetabular fractures is good. While high-energy mechanisms can cause unstable fractures and hemodynamic instability, pediatric patients tend to have less severe injuries, less hemorrhage, and less death compared to adults. Pelvic and acetabular fractures in skeletally mature patients tend to behave like adult fractures and should be treated as such. The pediatric surgeon should have a high suspicion for hemodynamic instability, associated head, visceral, and genitourinary trauma, and open fractures and initiate the appropriate management to optimize outcomes for these patients.

## Study Questions

A 4 year-old boy presents to the emergency department after being involved in a pedestrian versus motor vehicle accident. Per mother's report, he was able to walk after the injury. He is hemodynamically stable with no ecchymosis, open wounds, or open genitourinary trauma. He has tenderness over the posterior SI Joints, but is otherwise nontender throughout. Radiographs of the chest, abdomen, and pelvis are negative. What is the next best step?

A. Discharge the patient with close follow-up and instructions to return with any worsening symptoms.
B. Consult orthopedic surgery emergently.
C. Obtain CT pelvis.
D. Admit the patient for IV fluids and hemodynamic monitoring.

Answer: C.

*Explanation*: Plain radiographs can miss pelvic fractures up to 50% of the time. In this patient with high-energy trauma and positive examination findings (posterior SI tenderness), a CT pelvis should be ordered.

A 7 year-old girl presents to the emergency department after a motor vehicle accident. Pelvic binder was placed by the paramedics prior to arrival, and it is currently centered over the greater trochanters. She is found to have multiple displaced pelvic fractures on initial radiographs and CT imaging. There is no head trauma and no obvious visceral injuries. No other orthopedic injuries are identified. In the trauma bay, the patient becomes hemodynamically unstable despite ATLS resuscitation and massive transfusion protocol. What is the best next step?

A. Emergent OR with the pediatric surgery team for pelvic packing.
B. Emergent orthopedic and IR consultation.
C. Emergent transfer to the pediatric ICU in order to continue with the massive transfusion protocol.
D. Remove the pelvic binder and re-adjust to the appropriate position.

Answer: B.

*Explanation*: Persistent hemodynamic instability despite appropriate resuscitation measures require emergent external fixation with orthopedics to close down the pelvic volume and possible IR angiography/embolization. The pelvic binder is in the correct position, centered over the greater trochanters, and should not be readjusted.

## Further Reading

Banerjee S, Barry MJ, Paterson JM. Paediatric pelvic fractures: 10 years experience in a trauma centre. Injury. 2009;40(4):410–3. https://doi.org/10.1016/j.injury.2008.10.019. Epub 2009 Feb 20

Demetriades D, Karaiskakis M, Velmahos GC, Alo K, Murray J, Chan L. Pelvic fractures in pediatric and adult trauma patients: are they different injuries? J Trauma. 2003;54(6):1146–51; discussion 1151. https://doi.org/10.1097/01.TA.0000044352.00377.8F.

de la Calva C, Jover N, Alonso J, Salom M. Pediatric pelvic fractures and differences compared with the adult population. Pediatr Emerg Care. 2020;36(11):519–22. https://doi.org/10.1097/PEC.0000000000001411.

Guillaume JM, Pesenti S, Jouve JL, Launay F. Pelvic fractures in children (pelvic ring and acetabulum). Orthop Traumatol Surg Res. 2020;106(1S):S125–33. https://doi.org/10.1016/j.otsr.2019.05.017.

Miller MD, Thompson SR. Miller's review of orthopaedics. Philadelphia, PA: Elsevier; 2020.

Orthobullets.com. Pediatrics - Pediatric Trauma - Pelvis Fractures - Pediatric. 2021. Orthobullets.com, 2021 [online]. http://www.orthobullets.com.

Shaath MK, Koury KL, Gibson PD, Lelkes VM, Hwang JS, Ippolito JA, Adams MR, Sirkin MS, Reilly MC. Analysis of pelvic fracture pattern and overall orthopaedic injury burden in children sustaining pelvic fractures based on skeletal maturity. J Child Orthop. 2017;11(3):195–200. https://doi.org/10.1302/1863-2548.11.160266.

# Common Pediatric Fractures

**Brian K. Foster, Meagan Fernandez, and Louis C. Grandizio**

**Key Points**

1) Fractures are a significant cause of morbidity and mortality in the pediatric population.
2) Majority of pediatric fractures can be managed non-operatively.
3) Fractures with residual deformity can remodel over time.
4) Open fractures and compartment syndrome are surgical emergencies.
5) Non-accidental trauma needs to be considered in work-up (see non-accidental trauma chapter).

1. Anatomic considerations:
    (a) Physis
        i. aka growth plate.
            1. Complex organization of cartilage that create bone, leading to longitudinal growth.
            2. Radiolucent. Commonly mistaken for fracture lines.
            3. Close (fuse) in a consistent order and in predictable timeframes.

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    ii. Weaker than cortical bone which, pre-disposes to fracture.
        1. Ligamentous injuries are less common in pediatric patients.
  (b) Periosteum
    i. Thick, fibrous tissue that surrounds and supports bone.
    ii. Can aid in closed reduction of fractures.

2. Physeal injuries:
   (a) Fractures involving the growth plate can result in growth disturbance.
   (b) Salter-Harris classification: (Fig. 1)
      I. Fracture through the physis (widening of physis).
         a. Commonly missed
      II. Fracture through physis and metaphysis (most common).
      III. Fracture through physis and epiphysis.
      IV. Fracture through physis, metaphysis, and epiphysis.
      V. Crush injury to physis (narrowing of physis).
   (c) As a general rule, higher Salter-Harris grade results in increased incidence of growth disturbance.

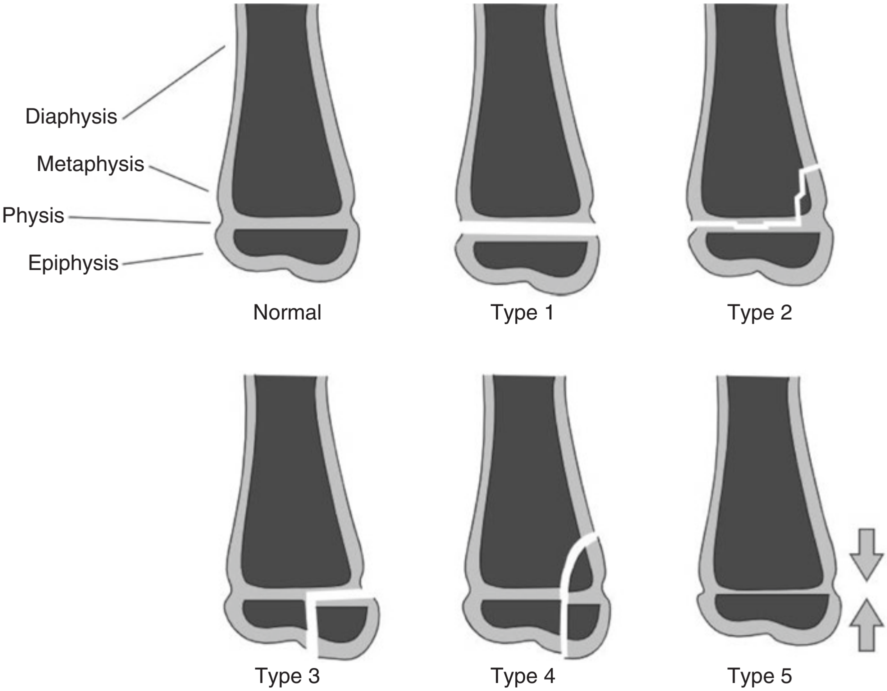

**Fig. 1** Salter-Harris classification of fractures. (Source: Christopher Coppola)

3. Deformity and remodeling:
    (a) In bones with open physes, fractured bones with residual deformity can remodel (straighten) over time.
        i. Pediatric fractures have greater remodeling potential than adult fractures.
        ii. Deformities in the plane of joint motion remodel best.
        iii. Rotation deformity does not reliably remodel.
    (b) Deformity is better tolerated closer to physis.
4. History:
    (a) Age, mechanism of injury, time of injury, witnesses to event, prior injuries.
    (b) Birth, developmental, family, and social history.
        i. Underlying metabolic bone disorders.
        ii. Osteogenesis imperfecta.
        iii. Risk factors for non-accidental trauma.
5. Physical examination:
    (a) All patients require a thorough, four extremity examination:
        i. In cases of distracting injuries, a later tertiary survey is required to assess for other injuries.
    (b) Skin:
        i. Any communication of fracture through skin is an open fracture.
        ii. Overlying lacerations and wounds may not occur at exact level of open fracture.
            1. Any overlying wound with continuous bloody "oozing" is assumed to be an open fracture.
        iii. "Impending open" fractures present with skin tenting from underlying bone.
            1. Can erode skin and convert closed fracture to open fracture.
            2. Requires emergent reduction.
        iv. Open fractures have higher infection risk. They require immediate IV antibiotics and urgent debridement in operating room.
            1. Tetanus booster status should be ascertained and administered, if out of date.
    (c) Soft tissues: assess myofascial compartments for soft-tissue swelling.
        i. Compartment syndrome:
            1. State of extreme swelling that compromises perfusion of distal limb.
            2. Can occur even without fracture (i.e. crush injuries, burns, bleeding).

        3. Compartments that are firm and not compressible are highly concerning.
        4. Assess for "5 P's":
            (a) Pain with passive stretch
            (b) Parasthesias
            (c) Pulselessness (late finding)
            (d) Paralysis
            (e) Poikilothermia (abnormal temperature)
        5. Examination can be unreliable in children.
            (a) "3 A's"
                i. Agitation,
                ii. Anxiety, and
                iii. increasing narcotic Analgesia requirement.
        6. Concern for compartment syndrome requires emergent orthopedic consult.
            (a) Treatment is fasciotomy in the operating room.
            (b) Missed compartment syndrome has devastating neurovascular and functional consequences.
    (d) Neurovascular examination:
        i. Complete neurologic examination:
            1. Motor and sensory examination within known nervous distributions.
            2. Can be limited by age and patient compliance.
                (a) Tip: try using toys to "play" with uncooperative children to conduct examination.
        ii. Vascular examination:
            1. Palpate peripheral pulses
            2. Doppler examination of pulses
            3. Assess perfusion of extremity
                (a) Capillary refill
                (b) Temperature
                (c) Color
6. Imaging:
    (a) X-rays
        i. Orthogonal views of the fracture are required.
        ii. Obtain radiographs of the joint above and below the fracture.
        iii. Comparative radiographs of contralateral extremity are helpful to distinguish fractures from normal anatomic variants.

      - iv. Descriptive terms:
        - 1. Location
          - (a) Physeal
          - (b) Metaphyseal
          - (c) Diaphyseal
        - 2. Complete vs incomplete
          - (a) That is, buckle (torus) or greenstick fracture
        - 3. Presence of intra-articular involvement
        - 4. Involvement of physis
        - 5. Angulation
        - 6. Displacement
        - 7. Comminution
  - (b) CT:
    - i. Consider for intra-articular fractures, physeal injuries.
    - ii. Increased exposure to radiation compared to plain radiographs.
      - 1. CT of the ankle is equivalent to 0.9 chest X-rays.
  - (c) MRI:
    - i. Limited use in the acute setting.
    - ii. Can be helpful to assess for physeal injuries and associated soft-tissue injuries.
  - (d) Ultrasound:
    - i. Use increasing secondary to availability and low cost compared to MRI.
    - ii. Useful to assess cartilage in unossified bone.

7. Initial treatment principles:
   - (a) Splinting:
     - i. Bony prominences and splint edges should be well to avoid skin and soft-tissue breakdown.
     - ii. Plaster or fiberglass (i.e. Orthoglass [BSN Medical, Hamburg, Germany]) can be utilized.
     - iii. Immobilization of fracture often require inclusion of joint above and/or below fracture.
     - iv. Need to immobilize fracture for pain control and maintenance of reduction.
       - 1. Splints should be appropriately molded to maintain reduction.
       - 2. Mantra: “curved splint makes a straight bone”.
     - v. Tight splints can lead to compartment syndrome.

        vi. Casts, which are circumferential, do not accommodate soft-tissue swelling and, therefore, are not used in the acute setting.
    (b) Reduction:
        i. Requires conscious sedation, intra-articular local anesthetic, or hematoma block depending on patient age and compliance.
        ii. Always obtain radiographs after reduction and splinting.

8. Common upper extremity fractures:
    (a) Clavicle fractures:
        i. Midshaft clavicle fractures are common.
        ii. Skin tenting at fracture site is a sign of impending open fracture.
        iii. Initial management of closed injuries is sling or shoulder immobilizer.
        iv. Majority can be managed non-operatively.
    (b) Proximal humerus fractures:
        i. Proximal humerus physis closes at age 14 in girls and 16 in boys.
        ii. Initial management with shoulder immobilizer or sling/swathe.
            1. Angulation better tolerated in patients <12 years old as they are able to remodel.
        iii. Most can be managed non-operatively.
            1. Unacceptable angulation treated with closed reduction and percutaneous pinning in the operating room.
    (c) Humerus shaft fractures:
        i. Most common long bone fracture seen in non-accidental trauma.
        ii. Holstein-Lewis fracture.
            1. Mid-distal 1/3 diaphyseal fracture associated with radial nerve palsy.
            2. Radial nerve palsy can be followed with serial examinations.
        iii. Initial treatment with sling and swathe for younger patients (<4 years old) or coaptation splint.
            1. Use additional wrist drop splint in presence of radial nerve palsy to avoid contracture.
        iv. Most can be managed non-operatively.
            1. Angulation up to 30 degrees is well tolerated.
    (d) Management of pediatric fracture about the elbow:
        i. Assessment of neurovascular status is critical.
        ii. Assess for increased compartment pressure.
            1. Missed compartment syndrome can result in Volkmann ischemic contracture with devastating functional outcomes.

            I. Emergent orthopedic consult and compartment pressure measurement with any concern for compartment syndrome.

    iii. Well-padded long-arm splints with the elbow at 70° of flexion are suitable for initial stabilization of most elbow fractures.

(e) Supracondylar humerus fractures:

    i. Most common pediatric elbow fracture
    ii. Classification:

        1. Extension type far more common than flexion type
        2. Gartland classification:

            I. Non-displaced
            II. Displaced with intact posterior cortex
            III. Complete displacement

    iii. Assess neurovascular status:

        1. Anterior interosseous nerve (AIN) palsy most common in extension-type fracture.
        2. Perfusion of hand is more important than presence of peripheral pulses.

            I. Cold, pulseless hand is a vascular emergency.

        3. Concerning examination findings include antecubital ecchymosis, “button-hole”, open fractures.
        4. Frequent monitoring for compartment syndrome.

            I. Increased risk of compartment syndrome with ipsilateral distal radius fracture.

    iv. Initial treatment principles:

        1. Splint in 70° of flexion with a well-padded long-arm splint.

            I. Assess pulses and perfusion after splint application.

        2. Type I fractures can be managed non-operatively.
        3. Type II and III fractures require closed vs. open reduction and percutaneous pinning.
        4. When there is risk for neurovascular compromise, absent radial pulses or poor perfusion, a closed reduction attempt should be performed by orthopedics in the emergency department under conscious sedation.

            I. Vascular compromise in the form of a cold, pulseless hand after reduction attempt requires emergent operative intervention and a vascular surgery consult.

(f) Forearm fractures:

  i. Overview:

    1. Can be classified as both bones forearm fractures, isolated radius or ulna fractures, Monteggia fractures, or Galeazzi fractures.
    2. Both bones forearm fractures can be complete or incomplete (greenstick fracture).
    3. Isolated fractures of the radius can be diaphyseal, metaphyseal (most common), or distal physeal.

      I. Closed vs. open treatment depends on patient age, fracture location, shortening, displacement, and angulation as well as neurovascular status.

    4. Monteggia fractures occur most commonly between ages 4 and 10 years old and involve a radial head dislocation with a fracture of the proximal ulna.

      I. Bado classification:

        I: Anterior radial head dislocation with apex anterior ulna fracture.
        II: Posterior radial head dislocation with apex posterior proximal ulna fracture.
        III: Lateral radial head dislocation with apex lateral proximal ulna fracture.
        IV: Anterior dislocation of the radial head with proximal radial and ulna fracture at the same level.

    5. Galeazzi fracture.

      I. Fracture of the distal radial diaphysis with disruption of the distal radiounlar joint (DRUJ).

        a. Often referred to as a “fracture of necessity” as it often necessitates operative treatment except in very young patients with anatomic reductions of the radius and the DRUJ.

  ii. Initial treatment principles:

    1. Both bones forearm fractures:

      I. Rule out neurovascular injury, associated fracture (floating elbow), and compartment syndrome.
      II. Initial splinting consists of a long-arm sugar-tong up, sugar-tong down splint.
      III. Avoid elbow flexion above 90°.

        a. Can “kink” vessels and potentially increase compartment pressures.

      IV. Orthopedic consult to determine need for closed reduction.
         a. Based on age, displacement, angulation, shortening.
   2. Isolated radius or ulna fracture:
      I. Rule out neurovascular injury, associated fractures, and compartment syndrome.
      II. Stable, incomplete fractures of the distal radial metaphysis without angulation can be initially managed with a short-arm well-padded splint or a sugar-tong splint around the elbow.
      III. Proximal or unstable fractures often require a closed reduction attempt by orthopedics as well as a long-arm sugar-tong up, sugar-tong down splint.
   3. Monteggia fracture:
      I. Require orthopedic consult.
      II. Bado types I–III can be managed with closed reduction of radial head and anatomic reduction of ulna fracture.
      III. Initial pre-reduction management is a long-arm sugar-tong up, sugar-tong down splint.
   4. Galeazzi fractures:
      I. Initial splinting includes sugar-tong splint around the elbow or long-arm sugar-tong up, sugar-tong down splint to immobilize the DRUJ.
      II. Adolescent patients or those without anatomic reduction of the radius and DRUJ often require operative intervention.

9. Common lower extremity fractures:
   (a) Femoral neck fractures:
      i. Femoral neck is the area between diaphysis and femoral head (in hip joint).
      ii. Rare fracture, caused by high-energy trauma.
         1. Assess for concomitant visceral and axial injuries and other fractures.
      iii. Orthopedic emergency.
      iv. Requires emergent surgery to stabilize restore blood flow to femoral head.
      v. High rate of complications, including avascular necrosis of femoral head, premature physeal closure, and leg length discrepancy.
   (b) Femoral shaft fractures:
      i. High suspicion for non-accidental trauma for patients <3 years old.
      ii. Treatment dictated by patient age, weight, and fracture pattern:
         1. <6 months: Pavlik harness

        2. 6 months–5 years: spica casting
        3. 5–11 years, length stable pattern: flexible intramedullary nails
        4. 5–11 years, length unstable: submuscular plate
        5. >11 years or near skeletal maturity: rigid intramedullary nail.

    iii. Fracture reliably heals, often with slight overgrowth.

(c) Fractures around the knee:

    i. Distal femur fracture.

        1. Fracture involving distal femoral physis.
        2. High incidence of growth arrest.
        3. Often requires surgery to achieve and maintain an adequate reduction.

    ii. Patellar sleeve fractures.

        1. Characterized by separation of cartilage "sleeve" from ossified patella.
        2. Commonly occurs in prepubescent males during athletic activity with large eccentric force across a flexed knee (i.e. landing from a jump).
        3. X-rays may not show fracture of patella, but instead a high-riding patella.
        4. Surgery is required to restore extensor mechanism.

    iii. Tibial spine fractures.

        1. Tibial spine is attachment for anterior cruciate ligament (ACL).
        2. High incidence of concomitant knee injuries (meniscus, cartilage).
        3. Initial treatment is immobilization in extension (splint vs knee immobilizer).
        4. Displaced fractures often require surgery to restore function of ACL.

    iv. Tibial tubercle fracture.

        1. Commonly occurs in adolescent males during athletic activity with large eccentric force across a flexed knee (i.e. landing from a jump).
        2. Beware of compartment syndrome.
        3. Fracture usually propagates along proximal tibia physis and into knee joint.
        4. Initial treatment is closed reduction and application of long leg splint.

            I. CT often used to evaluate intra-articular involvement.

        5. Surgery required to restore joint surface, except for non-displaced fractures.

(d) Tibial shaft fractures:

    i. May be the result of low-energy twisting injury or high-energy trauma.

        1. Beware of compartment syndrome in patients with high-energy mechanisms of injury.

- ii. Toddler's fracture: non-displaced spiral tibia fracture in children <2.5 that is commonly missed on initial X-rays.
    1. Requires immobilization and close follow-up.
    2. Classic story involves parents going down slide with child in lap.
- iii. Minimally displaced or incomplete fractures in younger children may be managed non-operatively with long leg immobilization.
- iv. Displaced fractures initially require closed reduction and placement of long leg splint.
    1. May require surgical stabilization, depending on fracture pattern, patient age, and concomitant injuries.
- v. Proximal fractures may develop a late valgus deformity that resolves spontaneously (Cozen's phenomenon).

(e) Ankle fractures:

- i. Triplane and Tillaux fractures.
    1. Characteristic fractures about distal tibia physis in children nearing skeletal maturity.
    2. Triplane: Salter-Harris IV fracture, seen in multiple planes.
        - I. Salter-Harris II on lateral X-ray and Salter-Harris III on AP X-ray.
    3. Tillaux: Salter-Harris III fracture of anterolateral epiphysis.
    4. Initial management is closed reduction and placement of long leg splint.
        - I. CT commonly used post-reduction to assess quality of reduction.
    5. May require surgery, depending on quality of reduction and age of patient.
- ii. Rotational ankle fractures.
    1. Involve combination of distal fibula (lateral malleolus) and/or medial malleolus.
    2. Most commonly due to supination-external rotation injuries.
    3. Initial management is closed reduction and splinting.
    4. May require surgery, depending on quality of reduction and age of patient.

## Study Questions

1. A 5-year-old girl sustains an injury of her left elbow after a fall off the monkey-bars. X-rays demonstrate a displaced extension-type supracondylar humerus fracture. On examination she has a palpable radial pulse but is not able to flex the

interphalangeal joint of the left thumb. What motor deficit is associated with the nerve most commonly injured in this fracture pattern?

1. Inability to extend the digits.
2. Inability to flex the interphalangeal joint of the thumb.
3. Inability to abduct the fingers.
4. Inability to extend the interphalangeal joint of the thumb.

Answer: (2). This most common nerve injured in an extension-type supracondylar humerus fracture is the anterior interosseous nerve (AIN). The AIN is a pure motor branch off the median nerve that innervates the flexor pollicis longus, flexor digitorum longus to the index and long fingers, and pronator quadratus. Injury to the AIN will result in the inability to flex the interphalangeal joint of the thumb and distal interphalangeal joints of the index and long fingers. Answer choice 1 and 4 are common findings in radial nerve palsy. Answer choice 3 is seen in ulnar nerve palsy.

2. A 16-month-old girl presents to the emergency department after an unwitnessed fall at home complaining of left leg pain. Radiographs demonstrate a diaphyseal femur fracture. In addition to stabilizing the fracture, what is the best next step in management?

    1. Skeletal survey and contacting child protective services.
    2. Emergent surgical intervention.
    3. Observation and discharge home.

Answer: (1). According to the American Academy of Orthopaedic Surgeons Clinical Practice Guidelines, there is strong evidence to support child abuse evaluation for children younger than 36 months with a diaphyseal femur fracture. Child abuse evaluation is often institutional dependent; however, this often includes skeletal survey and contacting children protective services.

3. Which sign is the most sensitive in the diagnosis of compartment syndrome in children?

    1. Pallor
    2. Paralysis
    3. Pulselessness
    4. Increasing analgesia requirement
    5. paresthesia

Answer: (4). Although the “5 P’s” (pain with passive stretch, paralysis, paresthesias, pulselessness, and pallor) are the more common signs and symptoms of compartment syndrome, these are often late findings. Bae et al., in a retrospective review of children with compartment syndrome, found that increasing analgesia frequency and dose requirement were early signs of compartment syndrome. In fact, only one patient in their study had all five of the “5 P” symptoms.

## Further Reading

American Academy of Orthopaedic Surgeons treatment of pediatric diaphyseal femur fractures evidence-based clinical practice guideline. https://www.aaos.org/globalassets/quality-and-practice-resources/pdff/pdffcpg.pdf. Published 5 Dec 5 2020.

Bae DS, Kadiyala RK, Waters PM. Acute compartment syndrome in children: contemporary diagnosis, treatment, and outcome. J Pediatr Orthop. 2001;21(5):680–8.

Hogue GD, Wilkins KE, Kim IS. Management of pediatric tibial shaft fractures. J Am Acad Orthop Surg. 2019 Oct 15;27(20):769–78.

Kocher MS, Sink EL, Blasier RD, et al. Treatment of pediatric diaphyseal femur fractures. J Am Acad Orthop Surg. 2009 Nov;17(11):718–25.

Livingston KS, Glotzbecker MP, Shore BJ. Pediatric acute compartment syndrome. J Am Acad Orthop Surg. 2017 May;25(5):358–64.

Olgun ZD, Maestre S. Management of pediatric ankle fractures. Curr Rev Musculoskelet Med. 2018;11(3):475–84.

Omid R, Choi PD, Skaggs DL. Supracondylar humeral fractures in children. J Bone Joint Surg Am. 2008 May;90(5):1121–32.

Pace JL. Pediatric and adolescent forearm fractures: current controversies and treatment recommendations. J Am Acad Orthop Surg. 2016 Nov;24(11):780–8.

# Part IV
# Neonatal Problems

# Fetal Surgery and Interventions

**Abigail J. Engwall-Gill and Erin E. Perrone**

**Key Points**

1. Fetal surgery is *in utero* intervention utilized to treat highly morbid or likely fatal congenital anomalies.
2. A multidisciplinary team is essential to ensure the accurate diagnosis, monitoring, and treatment of both patients, the fetus, and mother.
3. Tracheal occlusion via a balloon placed fetoscopically into the trachea is a treatment for lung hypoplasia associated with congenital diaphragmatic hernia.
4. Twin–twin transfusion syndrome occurs when vascular anastomoses connect the two twins in utero and result in exchange imbalances for both twins. One treatment option is to do fetoscopic laser ablation.
5. The first non-lethal indication for fetal surgery is myelomeningocele which is treated with open or fetoscopic prenatal surgery with defect coverage.

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1. Background

    a. Fetal surgery is a rapidly evolving, introspective field that has developed over the last seventy years from conceptual to multidisciplinary *in utero* interventions for a variety of diseases. Accurate diagnosis of a congenital anomaly that will have increased morbidity or mortality by awaiting delivery and postnatal management opens the door for consideration of fetal intervention. These interventions range from percutaneous, needle-based therapy to fetoscopic approaches to open fetal surgery. When offering fetal intervention, the disease pathophysiology and prognosis should be well established without effective or limited postnatal treatment options. Animal models are typically used to establish safety and understanding of fetal natural history prior to human studies. Fetal interventions should be performed only at fetal centers with expertise and all outcomes (positive and negative) must be reported, so as to continually improve and advance.
    b. A multidisciplinary team approach is crucial to this multifaceted and ethically complex field. This collaborative team includes at minimum: fetal sonologist, pediatric/fetal radiologist, pediatric/fetal surgeon, maternal-fetal medicine obstetrician, obstetric and pediatric/fetal anesthesiologists, pediatric/fetal cardiologist, geneticist, neonatologist, fetal nurse coordinator, and social worker.
    c. Imaging and diagnosis is crucial when discussing *in utero* interventions. Skilled sonographers and pediatric/fetal radiologists are essential in diagnosis and assessing viability of treatment. Prenatal ultrasound and fetal magnetic resonance imaging (MRI) are the basis of diagnosing and monitoring congenital anomalies.
    d. When discussing fetal conditions, one should understand fetal natural history, including hydrops, and the resultant implications. Hydrops is defined by fluid accumulation within two or more body cavities (pleural effusions, pericardial effusion, abdominal ascites, and/or skin edema). Fetal hydrops is a precursor of poor fetal and/or postnatal outcomes and often indicates severe fetal distress.
    e. Appropriate gestational timing for intervention is another important consideration. Depending on the disease process, the gestational age recommendations may change as many fetal interventions are completed in the second trimester and the risks and benefits of early delivery must be weighed against the risks and benefits of the intervention itself.

2. Needle-based procedures

    a. Cordocentesis is a diagnostic sampling of the umbilical vein via an ultrasound guided percutaneous approach. Fetal anemia can be diagnosed and treated via cordocentesis and *in utero* blood transfusions. Common causes of fetal anemia are isoimmunization (Rh incompatibility), parvovirus, and fetal–maternal hemorrhage. Cordocentesis can also be used to monitor and administer medications to the fetus.

   b. Fetal cardiac intervention (FCI)

      i. Aortic valvuloplasty is performed to avert progression of aortic stenosis to hypoplastic left heart syndrome (HLHS), which is an underdeveloped left heart incapable of supporting systemic circulation. Using ultrasound guidance, a needle is introduced *in utero* into the left ventricle and the narrowed aortic valve is balloon dilated.
      ii. Atrial septostomy is another percutaneous FCI used to treat HLHS in which the foramen ovale is found to be closed. This technique recreates a hole between the two atria allowing flow and encouraging growth in the left side of the heart.

   c. Shunts

      i. Vesicoamniotic shunt is used to create a connection between the obstructed urinary tract to the amniotic cavity. Bladder outlet obstruction can be caused by obstruction where the ureter connects to the kidney (ureteropelvic) or the bladder (ureterovesicular), but most commonly is caused by posterior urethral valves. Urinary obstruction can become lethal if resultant oligohydramnios leads to severe pulmonary hypoplasia.
      ii. Thoracoamniotic shunt is used to treat the potentially fatal accumulation of fluid in the thoracic cavity related to pleural effusions, congenital pulmonary airway malformations (CPAMS), and other compressive conditions.

3. Fetoscopic procedures are completed via endoscopic entry through the abdominal wall and uterus to allow access to the fetus, amniotic cavity, umbilical cord, and fetal placenta. There are multiple:

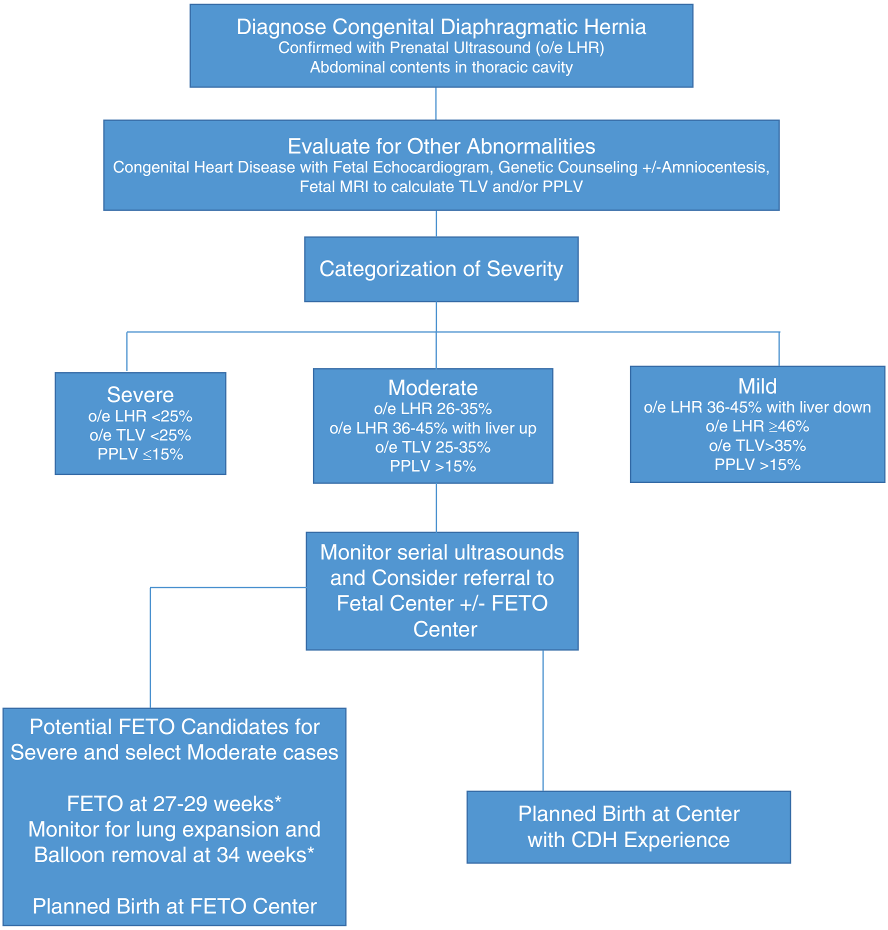
   a. Laser ablation is a fetoscopic technique used to ablate the aberrant placental connections between monochoriotic, diamniotic twins. Twin–twin transfusion syndrome (TTTS) is a blood exchange imbalance that causes the donor twin to become anemic with malnutrition and the recipient twin to become hypervolemic with cardiac complications. TTTS can additionally cause preterm labor. One treatment option is visualizing and ablating vascular anastomoses between the twins with a laser. Fetoscopic surgery has resulted in 70% survival of both twins in the last 20 years.
   b. Amniotic band ligation is considered if there is a recognized threatened limb due to concentric wrapping of amniotic bands around a fetal arm or leg. Treatment is fetoscopic ligation of the band to return blood flow to the affected limb but is only considered when this is identified early and the limb appears to be recoverable with intervention.
   c. Tracheal occlusion (TO) is another fetoscopic intervention and involves placing an obstructive balloon in the fetal trachea with the goal of promoting lung growth. Congenital diaphragmatic hernia (CDH) is a failure of the diaphragm to close allowing abdominal contents into the chest, which subsequently leads to pulmonary hypoplasia and pulmonary hypertension. The incidence

of CDH is 1 in 2200 and severity of disease is measured prenatally by a combination of ultrasound (US) and fetal magnetic resonance imaging (MRI) techniques by estimating fetal lung volumes. US measures the volume of lung on the unaffected side and compares it to the biparietal head circumference to adjust for gestational age. This gives a lung-to-head ratio (LHR) and observed-to-expected LHR (o/e LHR) which can be used to predict postnatal severity [1]. Fetal MRI is an adjunct to measure lung volume and is typically done in the second trimester in order to gain more information. Lung volumes are measured by either total lung volume (TLV) or percent predicted lung volume (PPLV); these measures are also used to predict postnatal severity [2, 3]. Postnatal management is the current standard of care; however, CDH remains a lethal anomaly in up to 30% of patients. Therefore, fetal intervention has been studied for the past 30 years with attempts to improve survival. Initially *in utero* therapy was aimed at diaphragmatic hernia repair via an open hysterotomy. However, the increased understanding of the pathophysiology and desire for more minimally invasive techniques progressed toward fetoscopic TO as this may cause accumulation of lung fluid and resultant lung growth. Not all patients are candidates for this procedure; thus, this is currently offered to those with poor prognostic factors for survival and is only done in select institutions. See Fig. 1 for an Algorithm for a proposed guideline for workup of prenatal CDH, categorization of severity based on known prenatal predictors, and consideration of fetal intervention.

d. Myelomeningocele (MMC) is an incomplete closure of the neural tube during development. This results in an exposed neural tissue and cerebral spinal fluid (CSF) leakage. Complications of untreated MMC are hindbrain herniation, hydrocephalus, cognitive disabilities, nerve incompetence resulting in bowel/bladder incontinence, and inability to walk. Hydrocephalus typically requires decompression with a ventriculoperitoneal (VP) shunt. The first damage of MCC anomaly is the abnormal neural placode and defect itself, which is made worse by the exposure to the amniotic fluid. Thus, it was proposed to perform *in utero* treatment by closing the neurotube defect. Recently, some centers have been performing this procedure fetoscopically in order to decrease the maternal risk; however, the nuances of fetoscopic versus open repair are beyond the scope of this chapter.

4. Open surgical procedures.

   a. Myelomeningocele (MMC) is the first non-lethal indication for fetal surgery as the morbidities previously discussed are functionally limiting and part of the morbidity is related to amniotic exposure. A randomized control trial was done to compare fetal repair to postnatal repair, entitled Management of Myelomeningocele Study (MOMS) [4]. The study demonstrated that prenatal repair was superior to postnatal repair as it resulted in decreased hindbrain herniation, reduced need for VP shunt at 1 year, and improved ability to walk without a device at 30 months. In this study, the MMC was repaired with an open approach, which requires an incision on the uterus (hysterotomy) and


Diagnose Congenital Diaphragmatic Hernia
Confirmed with Prenatal Ultrasound (o/e LHR)
Abdominal contents in thoracic cavity
Evaluate for Other Abnormalities
Congenital Heart Disease with Fetal Echocardiogram, Genetic Counseling +/-Amniocentesis, Fetal MRI to calculate TLV and/or PPLV
Categorization of Severity
Severe
o/e LHR <25%
o/e TLV <25%
PPLV ≤15%
Moderate
o/e LHR 26-35%
o/e LHR 36-45% with liver up
o/e TLV 25-35%
PPLV >15%
Mild
o/e LHR 36-45% with liver down
o/e LHR ≥46%
o/e TLV>35%
PPLV >15%
Monitor serial ultrasounds and Consider referral to Fetal Center +/- FETO Center
Potential FETO Candidates for Severe and select Moderate cases
FETO at 27-29 weeks*
Monitor for lung expansion and Balloon removal at 34 weeks*
Planned Birth at FETO Center
Planned Birth at Center with CDH Experience

*Timing may be different due to specific institutional/study protocols

**Fig. 1** An algorithm for treatment of Congenital Diaphragmatic Hernias. *CDH* congenital diaphragmatic hernia, *o/e* observed to expected, *LHR* lung to head ratio, *TLV* total lung volume, *PPLV* percent predicted lung volume, *FETO* fetoscopic endoluminal tracheal occlusion. Source: Abigail Engwall-Gill. Asterisk, Timing may be different due to specific institutional/study protocols

carries the burden of maternal risk which must be considered when discussing any potential intervention.

b. Teratomas are tumors derived from all three embryonic layers (ectoderm, mesoderm, and endoderm) and are generally benign. Teratomas that originate from the coccyx are called sacrococcygeal teratomas/tumors (SCT). The fetuses of concern are those which have a giant SCT as they are vulnerable to multiple complications ranging from preterm labor and intra-uterine fetal demise. The teratoma can act as a large arteriovenous fistula, which shunts blood from fetal circulation to the tumor which can cause high-output cardiac failure as well as non-immune hydrops. These large SCTs can lead to hemorrhage (internally or externally), anemia, hypovolemia, traumatic delivery causing hemorrhage, and difficult delivery due to the size distortion of the SCT. Poor prognosis associated with SCT are solid tumor morphology, fetal hydrops, and tumor volume to fetal weight ratio (TFR) greater than 0.12. Fetal intervention involves open fetal resection with debulking of tumor and preservation of neurologic structures; however, this is plagued with high fetal mortality.

c. Congenital pulmonary airway malformations (CPAMs) are rare multicystic masses, which replace fetal lung tissue. They are the most common congenital lung malformation and are connected to the tracheobronchial tree as well as the pulmonary vascular supply. The incidence is only 1 in 2000 live births. Antenatal ultrasound is the typical method of diagnosis with a minority of cases diagnosed in the neonatal period with CT scan for evaluating respiratory distress or recurrent infections. Poor prognostic indicators for CPAMs are fetal hydrops and CPAM volume ratio (CVR) >1.6. Once diagnosed, the fetus should be monitored with serial ultrasounds. Prenatal treatment for large CPAMs is initially managed with maternal steroids or thoracoamniotic shunting if there is a large dominant cyst (macrocystic). However, when the mass continues to grow or patient develops hydrops, a fetal resection versus delivery can be considered for microcystic lesions depending on the gestational age. Another consideration with CPAMs is the potential of maternal mirror syndrome where the mother develops pre-eclampsia (hypertension, vomiting, pulmonary edema, proteinuria, and/or placentomegaly). Mirror syndrome progression is life threatening to the mother and requires delivery.

5. Ex utero intrapartum therapy (EXIT) procedure

   a. An EXIT procedure is a specialized surgical delivery of the fetus where intervention can be done on the fetus prior to transition to postnatal circulation. General anesthesia is required for the mother and uterine relaxation is maintained. A hemostatic hysterotomy is created and the fetus is partially delivered but maintained on uteroplacental bypass for a period of time. While the fetus is oxygenated by the placenta, procedures may be done prior to delivery and conversion to neonatal circulation and oxygenation via the lungs. This is most commonly used for head and neck lesions that are thought to cause airway compromise. When left untreated, large neck masses have an 80–100% mortality. Different causes of large neck masses are cervical teratomas, lymphatic malformations, vascular malformations, thymic cysts, or neuroblastomas. Pathophysiology of a large neck mass can instigate esophageal obstruction with impaired swallow and tracheal obstruction; these blockades can lead to polyhydramnios, airway obstruction, and preterm labor. Indications for EXIT procedure are polyhydramnios, teratoma diagnosis, and tracheoesophageal displacement index (TEDI) greater than 12. TEDI was calculated by fetal MRI and the sum of the ventral and lateral displacement (in millimeters) from the ventral cervical spine.

b. Historically, EXIT to ECMO (extracorporeal membrane oxygenation) was a procedure trialed for CDH. This was offered with the hopes of improving CDH-associated life-threatening hypoxia, instability, and/or cardiac arrest in the time between delivery and ECMO. However, a survival benefit was not seen in those who received EXIT to ECMO verses those kids who were monitored postnatally with subsequent ECMO intervention. In EXIT to ECMO, the fetus's head and neck are delivered and cannulation for venoarterial ECMO is established before dividing the umbilical cord. EXIT to ECMO allows maintenance of fetal circulation while allowing opportunity for anatomical clarification; however, the indications are very limited at this time and not currently offered for the diagnosis of CDH.

c. Giant lung mass and cardiac tumors are two other diagnoses that are possible indications for EXIT procedure with or without ECMO. This is considered in extreme situations with the rationale for airway protection with mass resection on placental bypass.

d. Congenital high airway obstruction syndrome (CHAOS) is an upper airway obstruction which is associated with a poor prognosis if not identified prior to birth. The cause of intrinsic airway obstruction is most commonly laryngeal atresia but can also be laryngeal cysts, tracheal atresia, laryngeal/tracheal webs, subglottic stenosis/atresia, and tracheal/laryngeal agenesis. The pathophysiology of the airway obstruction (which led to the development of TO as a treatment used for prenatal CDH) leads to accumulation of lung fluid, which results in increased lung size, flattened diaphragm, and elevated intrathoracic pressures. A dangerous complication of CHAOS is that it can lead to compromised venous return and fetal hydrops. The EXIT procedure may be required to achieve a secure airway before delivery via intubation or tracheostomy. Postnatal airway management and reconstruction remains challenging in this patient population.

## Additional Notes

In 1961, Dr. William Liley performed the first invasive fetal procedure and thus conceived the inventive and introspective world of fetal surgery. He improved severe hydrops caused by severe anemia due to Rh incompatibility with a blind intra-abdominal blood transfusion. Dr. Michael Harrison, the recognized father of fetal surgery, started applying his understanding of fetal pathophysiology in 1981 by draining a bladder outlet obstruction with a pigtail shunt and continued investigations into multiple fetal diagnoses needing treatment, many of which are standard of care today.

Dr. Harrison was also one of the first to study open fetal surgery for repair of congenital diaphragmatic hernia (CDH). The initial studies showed that, in the more severe cohort of patients with liver in the chest, there was a high risk of fetal demise due to kinking of the umbilical vein after repair. In the less severe "liver down" group, although technically feasible, there was no survival advantage for fetal CDH repair over standard postnatal care. Subsequent studies involved open and fetoscopic clipping of the trachea followed by an EXIT procedure for tracheal clip removal as tracheal obstruction had shown increased lung volume in animal models of CDH. Currently, the procedure has evolved into fetoscopic endoluminal tracheal occlusion (FETO) and remains experimental.

Since its beginning, fetal surgery and intervention has grown immensely. This innovative field that set out to improve mortality and morbidity *in utero* now focuses on minimally invasive surgery with goals of reducing maternal risk while remaining focused on improving fetal outcome. Additionally, there are many collaboratives with clinical trials aimed at answering the question of prenatal versus postnatal therapy benefit. The question arises of when to refer expectant mothers to a fetal surgeon. The answer is whenever there is a concern for a potential anomaly that has a potential for prenatal therapy, an evaluation at a fetal center is recommended in order to give the patient the most up-to-date information.

This growing field has many exciting future directions. Placental stem cells are being used to augment the prenatal repair of MMC *in utero* with a neoskin, currently being studied by Dr. Diana Farmer and colleagues. Interestingly, fetuses have immature immune systems and do not require immunosuppressive therapy when receiving placental stem cells. Additionally, transamniotic stem cell therapy (TRASCET) has expanding applications and has been used in spina bifida as well as abdominal closure defects in animal models. TRASCET has shown decreased bowel inflammation in association with gastroschisis. Amniotic fluid-derived mesenchymal stem cells have been shown to home to placenta and fetal bone marrow, which suggests there may be a hematogenous spread rather than direct seeding.

Multicenter collaboration is important in fetal therapy given the rarity of the disease processes. The renal anhydramnios fetal therapy (RAFT) trial is aimed at addressing renal anhydramnios, which has an incidence of 1 in 2000 pregnancies. It is defined by different renal congenital diseases (bilateral renal agenesis, cystic kidney disease, and lower urinary tract obstructions) that result in anhydramnios by 22 weeks of gestation. These conditions are considered fatal due to anuria causing pulmonary hypoplasia, and perinatal palliative care or termination has been typical treatment option. Serial amnioinfusion has been trialed to replace the fluid in hopes to promote lung growth. The aim is that with increased lung function, the neonate may survive allowing their renal problems to be treated postnatally with renal replacement therapy and possibly transplant.

## Study Questions

1) What are indications for fetal intervention?
    a) Prevent mortality or major morbidity.
    b) To diagnosis a congenital anomaly.
    c) When there is a safe postnatal intervention.

Answer: a) Prevent mortality or major morbidity. The indication for fetal surgery is to prevent mortality or major morbidity. Fetal surgery should not be employed to diagnosis congenital anomalies, which can be done safely with ultrasound or fetal MRI. Additionally, surgical intervention during pregnancy should not be performed when there is a safe postnatal intervention with comparable results.

2) What is the name of the first randomized control trial to validate open fetal intervention compared to postnatal intervention in a non-fatal condition?

   a) Renal Anhydramnios Fetal Therapy
   b) Management of Myelomeningocele Study
   c) Ex Utero Intrapartum Therapy
   d) Transamniotic Stem Cell Therapy

Answer: b) Management of Myelomeningocele Study. The Management of Myelomeningocele Study (MOMS) is the first study to validate a non-lethal disease for fetal intervention. Myelomeningocele is the incomplete closure of the neural tube resulting in exposed neural tissue to caustic amniotic fluid as well as other neurologic complications. The study delineated multiple facets of improvement for those children treated *in utero* verses those who incurred more amniotic fluid exposure with a post-natal defect repair. Fetal intervention resulted in decreased hindbrain herniation, less VP shunts, and improved ability to walk without a device.

3) What are the current indications for EXIT (Ex-Utero Intrapartum Therapy) procedure?

   a) EXIT to ECMO for Congenital Diaphragmatic Hernia
   b) Congenital High Airway Obstruction Syndrome
   c) Twin–Twin Transfusion Syndrome
   d) Sacrococcygeal Teratomas

Answer: b) Congenital High Airway Obstruction Syndrome. Congenital High Airway Obstruction Syndrome (CHAOS) is an indication for EXIT procedure where a safe airway is established before ligating placental bypass and delivering the baby. Large neck masses are another potential indication for EXIT especially when associated with polyhydramnios, teratoma diagnosis, and/or TEDI >12. Although trialed in the past, CDH is no longer typically treated with EXIT to ECMO. Twin–twin transfusion syndrome is treated with fetoscopic laser ablation of aberrant vessels. Giant SCTs with impending hydrops, solid tumor morphology, or TFR > 0.12 are recommended to undergo fetal debulking surgery if less than 27 weeks gestation.

## References

1. Deprest JA, Flemmer AW, Gratacos E, Nicolaides K. Antenatal prediction of lung volume and in-utero treatment by fetal endoscopic tracheal occlusion in severe isolated congenital diaphragmatic hernia. Semin Fetal Neonatal Med. 2009;14(1):8–13.

2. Oluyomi-Obi T, Kuret V, Puligandla P, et al. Antenatal predictors of outcome in prenatally diagnosed congenital diaphragmatic hernia (CDH). J Pediatr Surg. 2017;52(5):881–8.
3. Barnewolt CE, Kunisaki SM, Fauza DO, Nemes LP, Estroff JA, Jennings RW. Percent predicted lung volumes as measured on fetal magnetic resonance imaging: a useful biometric parameter for risk stratification in congenital diaphragmatic hernia. J Pediatr Surg. 2007;42(1):193–7.
4. Adzick NS, Thom EA, Spong CY, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. N Engl J Med. 2011;364(11):993–1004.
5. Fetal Diagnosis and therapy: a reference handbook for pediatric surgeons, 2nd ed. Electronic Publication of the American Pediatric Surgical Association, 2019.
6. Jancelewicz T, Harrison MR. A history of fetal surgery. Clin Perinatol. 2009;36:227–36.
7. Deprest JA, Flake AW, Gratacos E, Ville Y, Hecher K, Nicolaides K, Johnson MP, Luks FI, Adzick NS, Harrison MR. The making of fetal surgery. Prenat Diagn. 2010;30:653–67.
8. Johnson MP, Bennett KA, Rand L, et al. The management of myelomeningocele study: obstetrical outcomes and risk factors for obstetrical complications following prenatal surgery. Am J Obstet Gynecol. 2016, 215:778.e1–9.
9. Perrone EE, Deprest JA. Fetal endoscopic tracheal occlusion for congenital diaphragmatic hernia: a narrative review of the history, current practice, and future directions. Transl Pediatr. 2020;10(5):1448–60.

# Necrotizing Enterocolitis

**Samir Narula, Pratibha Vemulapalli, and Brian Gilchrist**

**Key Points**

1) Necrotizing enterocolitis (NEC) is the most common emergency encountered by pediatric surgeons in the neonatal intensive care unit (NICU). It is an acquired inflammatory disease affecting the immature gastrointestinal tract of premature infants.
2) NEC is multifactorial and incompletely understood but classically occurs after an insult results in mucosal compromise of the gut barrier in infants receiving enteral feeds that leads to an exaggerated immune response that can ultimately result in intestinal necrosis.
3) Although management is initially supportive with fluid resuscitation, gastric decompression, broad spectrum antibiotics, and total parenteral nutrition, up to 50% of infants will require operative intervention in the acute stage.
4) There is ongoing debate regarding whether peritoneal drainage (PD) or laparotomy is the treatment of choice for perforated NEC in extremely low-birth-weight neonates.

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1. Background

   Necrotizing enterocolitis (NEC) is an acquired inflammatory disease that affects the gastrointestinal tract primarily in premature infants. Although the term *necrotizing enterocolitis* was not coined until the 1950s, reports of intestinal necrosis in neonates date back to the 1800s. We are yet to fully understand the pathogenesis, which is multifactorial. However, there is a consensus that it involves a combination of intestinal mucosal damage, intestinal dysbiosis, and enteral feedings which leads to progressive enteric mucosal injury and inflammation.
2. Epidemiology
   1. Incidence:

      NEC has proven to be difficult to eradicate and persists as the most common emergency encountered by pediatric surgeons in the NICU. The overall incidence is 1–3 cases per 1000 live births, accounting for 1–7% of all NICU admissions in the United States. The degree of prematurity and low birth weight are important factors in developing NEC, with over 90% of affected infants being premature and weighing less than 2000 grams (g).
   2. Risk factors:

      Numerous risk factors are associated with the pathogenesis of NEC. The incidence of NEC has consistently shown an inverse relationship to birth weight and gestational age, with the highest incidence in those weighing <750 g (14%). Intestinal dysbiosis, defined as an imbalance of the gut microbiome, allows for overgrowth of non-commensal bacteria and increases the risk for NEC. The use of antacids and antibiotics in the NICU is associated with dysbiosis. The initiation of formula feeds also plays a role, with approximately 90% of cases developing after feedings are initiated. It is also associated with increased volume and osmolality of feeds. Breast-fed infants are colonized by mostly gram-positive organisms which help prevent the overgrowth of the gram-negative bacteria, which are commonly found in formula-fed infants. Maternal factors such as chorioamnionitis, cocaine abuse, in utero growth restriction, and many others have inconsistently been implicated. The prolonged use of indomethacin, acute hypoxia, anemia, and cardiac anomalies among others has also been reported.
3. Pathophysiology and pathogenesis

   The pathogenesis of NEC is multifactorial and yet to be fully elucidated. The prevailing understanding of classic NEC which occurs in premature neonates includes an immature gastrointestinal (GI) tract and intestinal barrier, an event leading to mucosal compromise, an exaggerated immune response to non-commensal (often gram-negative) organisms that have colonized the gut, and the presence of enteral feeds in the gut (most commonly formula). These factors together cause overwhelming levels of inflammation which lead to progressive tissue injury, increased intestinal permeability, and potentially necrosis.

   The initial insult is hypothesized to be hypoxia, poor perfusion, or infection. The damaged mucosa is then invaded by intraluminal bacteria which activates an intensifying immune response leading to further damage. Gram-negative bacteria have the endotoxin lipopolysaccharide (LPS) which is a potent activator of inflammatory response and has been found in high quantities in NEC. The bacteria induce damage to and inhibit repair of the epithelium by promoting the release of substances such as nitric oxide, interferon gamma, and interleukins. Interactions between these and other cytokines, inflammatory mediators, and immune effectors lead to breakdown of the gut barrier and bacterial translocation. This results in an overwhelming inflammatory response which induces apoptosis, localized vasoconstriction, as well as activation of the coagulation and complement systems. These complex interactions between enterocytes, bacteria, and the immune system culminate in intestinal necrosis. Interestingly, NEC may involve one or more segments of the GI tract. It is most frequently seen in the terminal ileum, with around 40% of cases affecting both the small and the large intestine.

Spontaneous intestinal perforation (SIP) is a unique type of perforation that occurs in the terminal ileum generally on the antimesenteric border which is not associated with obvious necrotizing enterocolitis. It is believed to be the result of compromised blood flow of the watershed area of the ileocolic artery. SIP generally occurs during the first week after birth and is seen in more mature or term neonates than in NEC. While clinically the presentation of SIP appears NEC-like, this condition is associated with minimal intestinal inflammation and necrosis which is often reflected by subtler levels of inflammatory markers. No known association with enteral feeding exists, rather it is related to other problems. It has been associated with medications which cause vasoconstriction of the splanchnic circulation in particular indomethacin (which is administered for patent ductus arteriosus). It is also more frequently associated with maternal drug use, congenital heart disease, and any perinatal stressor that affects mesenteric blood flow.

4. Clinical features

   The early clinical presentation of NEC is often nonspecific, reflecting a state of evolving physiologic instability. The findings include recurrent apnea, bradycardia, lethargy, temperature instability, or increased metabolic requirements. More specific clinical findings may be present, including feeding intolerance (reflected by emesis or high gastric residuals), abdominal distention, and blood per rectum. The increase in metabolic requirements and abdominal distention can result in a sudden rise in ventilatory requirements.

5. Diagnosis

   a. Symptoms:

      While the initial presentation of NEC is often subtle and nonspecific, certain GI tract symptoms may be present. The most common findings in the order of decreasing incidence include abdominal distention, blood per rectum, high gastric residuals, emesis, and diarrhea. Flecks of gross blood in stool or occult bleeding are common, with massive bleeding seen rarely.

b. Examination:

Initial physical examination may only reveal subtle abdominal distention (70–98%) with minimal tenderness. As NEC progresses, palpation of the abdomen can provoke a considerable amount of tenderness, increasing rigidity, palpable loops of bowel, a fixed or mobile mass, or crepitus. Sometimes, skin color changes of the abdominal wall are observed. If duskiness is appreciated, it signifies adjacent discoloration of the bowel wall or feculent peritonitis seen through the neonate's thin abdominal wall. Erythema and edema of the abdominal wall suggest peritonitis and are generally appreciated late in the course; however, they are rarely present on initial assessment. In males, it is prudent to examine the scrotum for discoloration suggestive of perforation. While only seen in 10% of NEC cases, a fixed abdominal mass (suggesting a non-peristaltic, necrotic loop of bowel) and erythema of the abdominal wall are concerning signs and a relative indication for surgical intervention, though not absolute.

c. Laboratory tests:

Conventional laboratory tests aid in ascertaining the degree of systemic illness imposed on the neonate. Either leukocytosis with bandemia or leukopenia may be seen, with the latter often indicating more severe illness. Neutropenia (<6000 cells/mm$^3$) is commonly seen in those with gram-negative septicemia. Thrombocytopenia is commonly present (65–90%) and is associated with gram-negative sepsis and binding of platelets by the endotoxin LPS. Multiple reports have demonstrated that a platelet count of <150,000 cells/mm$^3$ corresponds to the degree of severity of NEC and is commonly present in neonates warranting surgical intervention. Precipitously worsening thrombocytopenia and count less than 10,000 cells/mm$^3$ have been shown to be poor prognostic indictors. Metabolic acidosis is present in 40%–85% of NEC patients reflecting a compromised state of perfusion due to hypovolemia and sepsis.

Numerous biomarkers have been evaluated for their utility in early diagnosis of NEC and to stratify those who may benefit from operative intervention before evidence of perforation exists. Acute phase reactants, notably C-reactive protein (CRP), and various interleukins have been extensively studied to determine levels predicting presence, progression, or resolution of NEC. An increase in CRP levels, >10 mg/L within 48 h of suspected diagnosis, may be an early indication of NEC, and failure to normalize within 10 days suggests abscess, stricture, or septicemia. While relatively sensitive (92%) for NEC, CRP fails to have significant specificity.

Fecal calprotectin (found in neutrophils and macrophages) is a well-established marker of intestinal inflammation, and elevated levels are associated with perforated NEC. However, inconsistency in levels and difficulty in collecting stool specimens in neonates limit the diagnostic value.

Another marker is intestinal fatty acid binding protein (I-FABP), originally from enterocytes of small bowel villi. I-FABP is released into circulation and excreted renally following intestinal damage and enterocyte death. High urinary concentration has been seen in those requiring surgical intervention and is associated with poor prognosis.

Various biomarkers may provide insight into the severity or prognosis of NEC; however, presently their use as a diagnostic tool is limited. There is an ongoing global effort to find clinically useful biomarkers to better identify those at risk for NEC and to better predict which neonates warrant operative intervention before perforation. The results of current research may provide the tools necessary to improve NEC outcomes for the first time in decades.

d. Radiographic studies:

The overall clinical presentation of NEC, except abdominal rigidity and skin changes, is nonspecific. Thus, imaging is often the critical piece of information yielding the diagnosis of NEC. Initial evaluation is generally made using plain anteroposterior and left lateral decubitus radiographs; identification of pneumatosis intestinalis (PI) is pathognomonic for NEC and present in 70–80% of cases. Pneumatosis intestinalis may either be submucosal giving the linear appearance of air in the bowel wall or the "soap suds" appearance of subserosal air which may be difficult to distinguish from feces within the bowel. However, various radiographic findings with different clinical implications may be seen in patients suspected to have NEC. Such findings include nonspecific bowel distention (ileus pattern), air-fluid levels, portal venous gas, pneumoperitoneum, intraperitoneal free fluid, and persistently dilated fix loop(s) of bowel. The presence of portal venous gas is a poor prognostic sign. The presence of pneumoperitoneum and intraperitoneal free fluid suggest perforation.

Ultrasonography (US) has been shown to be effective in detection of bowel peristalsis, wall thickening, reduction of perfusion (using Doppler mode), free fluid, pneumatosis intestinalis, portal venous gas, and pneumoperitoneum. Some studies have suggested that US is more sensitive than plain film radiographs in diagnosing NEC due to its ability to detect more subtle findings such as fluid and air. However, due to lack of large prospective studies, the use of ultrasound is not widespread and requires further study.

6. Treatment

a. Initial measures:

Without evidence of intestinal necrosis or perforation on presentation, cases of NEC are managed supportively with bowel rest, gastric decompression, broad spectrum intravenous (IV) antibiotics, IV fluid resuscitation, and parenteral nutrition. Cardiopulmonary support is frequently warranted. Since no regimen shows superiority, antibiotic selection should be guided by the local resistance patterns and provide coverage for gram-positive, gram-negative, and anaerobic organisms. Antifungal therapy should be considered in neonates with a history of prolonged antimicrobial therapy. Thrombocytopenia may signify a fungal infection. A commonly used regimen includes ampicillin, an aminoglycoside (gentamicin) or third-generation cephalosporin (cefotaxime), and clindamycin or metronidazole. Piperacillin-tazobactam may also be used. Fluconazole is often added for prophylaxis against fungal overgrowth. The duration of treatment is based on the Bell's stage (Fig. 1).

b. Subsequent measures:

Once medical management is initiated for suspected (Bell's stage 1) NEC, close clinical and radiographic evaluation by the neonatology and surgical teams for operative indications is crucial. This generally includes frequent abdominal examinations, two-view abdominal radiographs, ultrasonography, complete blood counts, and blood gas analysis. If pneumatosis intestinalis does not develop in the subsequent 72 h, treatment can be discontinued. If progression to Bell's stage 2 occurs, medical management is continued for 7–10 days with ongoing reevaluation for operative indications. Enteral feeds may be resumed slowly in those who improve with medical management. Once feeds are resumed, the infant should be monitored for signs or symp-

| Stage | Signs | Abdomen | X-ray | Treatment |
|---|---|---|---|---|
| **I Suspected** | | | | |
| IA | Temperature instability, apnea, bradycardia, lethargy | Gastric distention, emesis, heme-positive stool | Normal or intestinal dilation, mild ileus | NPO, 3 days antibiotics |
| IB | Same as above | Bloody stool | Dilation or ileus | NPO, 3 days antibiotics |
| **II Define** | | | | |
| IIA Mildly ill | Same as above | Same as above, absent bowel sounds, +/- tenderness | Dilation, ileus, pneumatosis intestinalis | NPO, 7-10 days antibiotics |
| IIB Moderately ill | Same as above, metabolic acidosis, thrombocytopenia | Same as above, no bowel sounds, tenderness, +/- cellulitis, mass | Same as above, ascites | NPO, 14 days antibiotics |
| **III Advanced** | | | | |
| IIIA Intact Bowel | Same as above, shock, bradycardia, apnea, respiratory acidosis, DIC, neutropenia | Same as above, peritonitis, tenderness, distention | Same as above | NPO, 14 days antibiotics,<br>IV fluid,<br>pressors,<br>ventilator therapy,<br>paracentesis |
| IIIB Perforation | Same as above | Same as above | Same as above, free air | Laparotomy |

**Fig. 1** Modified Bell's staging criteria for necrotizing enterocolitis. (Adapted from: Coppola CP. Necrotizing enterocolitis. In: Coppola CP, Kennedy AP, Scorpio RJ, editors. Pediatric surgery: diagnosis and treatment. Springer, 2014:123–126)

toms of NEC and stool tested for reducing substances or occult blood. Any positive findings necessitate prompt discontinuation of feeds. Patients with advanced NEC (Bell's stage 3) who fail to improve despite maximal medical therapy or develop pneumoperitoneum require surgical intervention.

While there are currently no absolute indications for paracentesis, some studies recommend its use in patients with extensive pneumatosis or who do not improve with conservative management. A positive result is highly specific for intestinal necrosis and is defined as free-flowing aspiration of more than 0.5 mL of brown or yellow-brown fluid that contains bacteria on gram stain. If no fluid is aspirated, peritoneal lavage is performed, and the patient is turned from side to side, then the fluid is withdrawn and analyzed. A negative result in the presence of necrosis may occur if walled-off perforation is present.

Prior to proceeding to the operating room, the infant should be optimized without delaying intervention. This may include aggressive ventilatory support, treatment of shock (inotropes, vasopressors, and/or fluid resuscitation), broad spectrum antibiotics, correction of anemia, and coagulopathy. It is also important to ensure that blood products are readily available intra- and postoperatively.

c. Operative treatment:

The presence of pneumoperitoneum, positive paracentesis, and clinical deterioration despite maximal medical therapy are absolute indications for operative intervention. Relative indications are controversial and include a palpable abdominal mass, abdominal wall erythema, portal venous gas, and fixed intestinal loop (suggestive of necrosis). In practice, the decision to operate is made based on the individual surgeon's clinical judgment and previous experience.

Primary peritoneal drainage (PPD) was initially developed as a temporizing measure in which Penrose drains are inserted in unilateral or bilateral lower quadrants in the most unstable and premature neonates as a bridge to laparotomy. Attempts to use PPD for definitive treatment have occurred since a report published in the 1970s showed some patients never required laparotomy after undergoing drainage. It is generally performed in premature infants weighing <1000 g and is done as a bedside procedure. It is likely most successful in premature infants with spontaneous intestinal perforation.

During the acute stage of the disease, up to 50% of infants will eventually require operative management (Bell's stage 3b). The objective of surgical intervention for NEC is removal of gangrenous bowel, preservation of intestinal length, and to control sepsis. The ideal procedure performed is based on the extent of intestinal involvement seen during laparotomy, as well as the patient's overall condition as illustrated in Fig. 2. Involvement of segments of both the small and the large intestine is commonly found. The degree of involvement is classified as focal (one segment), multifocal (more than one segment affected), or pan-intestinal. It is essential to resect only frankly necrotic bowel to maintain as much length as possible.

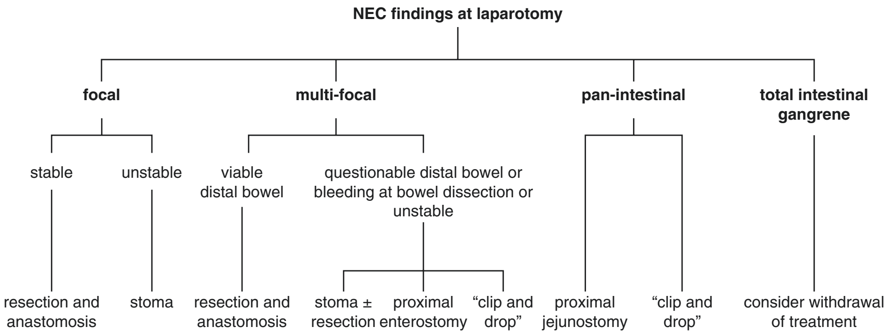
NEC findings at laparotomy
focal
multi-focal
pan-intestinal
total intestinal gangrene
stable
unstable
viable distal bowel
questionable distal bowel or bleeding at bowel dissection or unstable
resection and anastomosis
stoma
resection and anastomosis
stoma ± resection
proximal enterostomy
“clip and drop”
proximal jejunostomy
“clip and drop”
consider withdrawal of treatment

**Fig. 2** Surgical decision making for NEC. (Adapted from: Pierro A, Hall N. Surgical treatment of infants with necrotizing enterocolitis. Semin Neonatol. 2003;8(3):223–232. https://doi.org/10.1016/S1084-2756(03)00025-3)

Resection of necrotic bowel and proximal stoma and distal mucus fistula creation is the traditional procedure of choice when focal disease in encountered. While rare, resection and primary anastomosis can be considered for isolated disease in select patients to avoid the complications associated with a proximal stoma (high output, electrolyte imbalances, and malnutrition). This is no universally agreed upon time for restoration of intestinal continuity. Factors to consider before reversal include time since surgery (>4 weeks), weight gain, stoma output, and the need for or effects of total parenteral nutrition (TPN) on metabolic function. It is important to perform a contrast enema to assess for strictures prior to restoration of continuity.

Multifocal disease is defined by multiple areas of necrotic bowel separated by healthy bowel with >50% viable intestine. The goal of surgery in these patients is to preserve as much length of bowel as possible. Many of these operative approaches will require a second operation. Surgical options when multifocal NEC is encountered include:

- Each diseased segment resected, and multiple stomas created.
- Resection of diseased areas and multiple anastomoses to preserve bowel length.
- “Skewer”—proximal jejunostomy is created and the distal ends of bowel spliced together.
- “Patch, drain, and wait”—approximate any perforations using suture, insert two Penrose drains that exit in the lower quadrants, and start long-term TPN.
- “Clip and drop”—resect the areas of necrotic bowel, close the cut ends with staples or clips, re-explore in 48–72 h, assess need for further resection, remove all staples/clips, and anastomose all segments.

Pan-intestinal NEC (>75% of small and large bowel) poses a challenge, as removal of all necrotic areas increases the subsequent risk of short gut syndrome, while failure to resect these areas will lead to worsening sepsis. If there is doubt about the viability of some areas, the “clip and drop” or “patch, drain, and wait” techniques with a proximal jejunostomy may be performed, with a second look laparotomy after stabilization and weight gain. Irrespective of the approach chosen, the mortality rate in these cases is extremely high. Finding of NEC totalis (<25% viable bowel) should prompt closure of the abdomen and an honest discussion with the family regarding withdrawal of care.

d. Postoperative treatment:

A systemic inflammatory response will likely occur postoperatively, and support of failing organ systems is often needed. IV antibiotics are continued for 1–2 weeks, and feeds re-introduced once the infant has stabilized and bowel function returns.

e. Potential complications:

- Recurrence—Rates in the literature vary but have been reported as high at 10%, more likely to occur in those with other congenital anomalies and of lower birth weight.
- Strictures—May develop in those treated medically or surgically, although more often in those with surgery (up to 50%). They can occur anywhere but most commonly in watershed areas of the colon (most common: descending colon). They can be silent and found on imaging, present as partial or intermittent bowel obstructions and rarely with perforation. Treatment is laparotomy, resection of strictured area and primary anastomosis; in select cases, endoscopic dilation can be used.
- Intestinal failure/short gut syndrome—Inability to meet nutritional and fluid requirements, may be due to length or functionality, require TPN. The estimated incidence is as high as 23% in those that survive. Intestinal failure occurs in medical and surgical NEC, but more common in surgical.
- Neurodevelopmental impairment—Affects half of survivors; surgical NEC is at highest risk, incompletely understood at this time but has been speculated that white matter injury occurs due to hemodynamic instability and cytokine exposure.
- Stoma complications—Wound infections and dehiscence, stoma stenosis and prolapse, parastomal hernia (may cause small bowel obstruction (SBO)), and dehydration due to high output.
- Mortality—Rate nears 30% for medical NEC and up to 50% for surgical NEC. Despite intensive ongoing efforts to further understand NEC and improve outcomes, morbidity and mortality rates have remained unchanged for decades. Mortality and morbidity is inversely proportional to birthweight and gestational age. The mortality rate is higher in those with congenital anomalies, particularly of the heart. Mortality secondary to intestinal failure is affected by location of disease with more proximal small bowel having the worst outcomes and isolated colonic disease having the best.

## Additional Notes

Since its discovery, standard management has shifted from early operations to supportive medical care unless evidence of perforation is present. Despite being first reported in France two centuries ago, NEC continues to be poorly understood, devastating, and a common disease encountered in the NICU. In recent decades, the world has seen increasing rates of preterm birth as well as dramatic improvements in neonatal intensive care allowing the youngest and smallest neonates to survive, thus the incidence of NEC has also increased. The incidence of NEC varies worldwide, with expected variations seen between urban and rural communities as well as high- and low-income countries owed to the resources available.

In 1977, the first report of primary peritoneal drainage by Marshall and Ein in Toronto was published. Normally intestinal perforation is treated with laparotomy, resection, and ostomy. The authors described a last-ditch effort at survival performed bedside under local anesthesia on five small, very premature, and sick neonates that would likely not tolerate laparotomy. While too sick to operate on, the chance of survival was negligible without evacuation of the air, pus, and stool that had accumulated intra-abdominally. Although post-drainage all neonate's conditions remained poor and prognoses grim, they began to improve clinically within days, and all had bowel movements per rectum within 1 week. They were closely monitored after drainage for signs of further deterioration requiring repeat drainage or a formal operation. None of the babies required laparotomy after drainage, and only one required repeat drainage. Ironically, three of the babies survived. Two succumbed to other causes but did so with functioning and intact GI tracts.

The role of PPD, if any, in managing neonates with perforated NEC has long been debated and to date remains unclear and controversial. Dozens of studies including retrospective studies, systematic reviews, meta-analyses, and several multi-center prospective trials have sought the answer to the question, with unclear results. Some have shown higher mortality in the PD group, while others have shown no difference in outcome between groups. Varying rates in the need for laparotomy after PD have also been reported. In the time since 1977, it has been recognized that SIP is a separate and distinct disease process. However, many preceding studies likely included both SIP and NEC patients, explaining why at least half in most series on PPD for perforated NEC did not require additional surgery. It has since been shown that PD is more likely to allow the perforation to seal in SIP and may be the optimal procedure in these patients.

In summary, some still suggest PPD as definitive therapy, while most feel it should only be used as an initial temporizing measure to control sepsis and bridge the smallest and sickest to definitive laparotomy as shown in Fig. 3. This feeling was shared by Ein in his follow-up paper published 13 years after his initial paper. Given the mixed results of various publications on the role of PPD in perforated NEC since being first introduced, to no surprise there is little consensus concerning ideal surgical management. Recently, the Necrotizing Enterocolitis Surgical Trial (NEST)

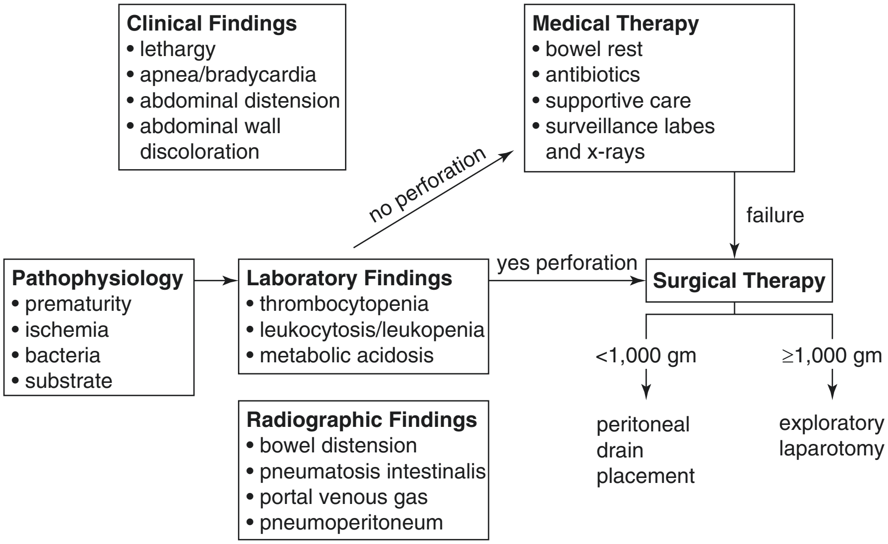
Clinical Findings
• lethargy
• apnea/bradycardia
• abdominal distension
• abdominal wall discoloration
Medical Therapy
• bowel rest
• antibiotics
• supportive care
• surveillance labes and x-rays
no perforation
failure
Pathophysiology
• prematurity
• ischemia
• bacteria
• substrate
Laboratory Findings
• thrombocytopenia
• leukocytosis/leukopenia
• metabolic acidosis
yes perforation
Surgical Therapy
<1,000 gm
≥1,000 gm
peritoneal drain placement
exploratory laparotomy
Radiographic Findings
• bowel distension
• pneumatosis intestinalis
• portal venous gas
• pneumoperitoneum

**Fig. 3** Treatment algorithm for necrotizing enterocolitis. (Adapted from: Glynn LA. Necrotizing enterocolitis. In: Saclarides TJ, Myers JA, Millikan KW, editors. Common surgical diseases an algorithmic approach to problem solving. 3rd ed. Springer, 2015:299–301:chap 76)

completed; this long-term multi-center study randomizing extremely low birth weight infant (ELBW) infants with perforated NEC to PPD vs. laparotomy is expected to help surgeons make informed decisions regarding the use of PPD. The author expects to find lower rates of mortality and long-term neurodevelopment impairment in those who undergo initial laparotomy rather than PPD.

## Study Questions

1) Overnight you are called to evaluate a 2-week-old premature baby girl with a birthweight of 900 g, who was being fed formula that begins to experience episodes of apnea and persistent bradycardia and ultimately required intubation. Examination reveals mild abdominal distention and tenderness. An abdominal X-ray shows uniform distention of the bowel with gas in the rectum, no pneumatosis intestinalis, and no evidence of pneumoperitoneum. Development of which of the following while being treated medically is an absolute indication for surgical intervention?

   A. Thrombocytopenia.
   B. Fixed loops on plain abdominal film.
   C. A palpable abdominal mass.
   D. Pneumoperitoneum on abdominal X-ray.

Answer to Question 1:

(D) The only absolute indication for surgical intervention in necrotizing enterocolitis is the presence of free air seen on abdominal imaging. Given the low birth weight of the infant, the intervention of choice may be primary peritoneal drainage followed by laparotomy if no improvement. Choices B and C are only relative indications for surgery. Choice A is part of Bell's modified criteria Stage 2B—Definite NEC and should raise suspicion for bowel perforation however not an indication for surgery.

2) A premature infant born at 32 weeks' gestational who was previous feeding on formula is found to have abdominal distention, tenderness, and erythema. Stool is positive for fecal occult blood. Laboratory studies show neutropenia, thrombocytopenia, metabolic acidosis, and gram-negative bacteremia. Abdominal X-ray shows pneumatosis intestinalis without evidence of pneumoperitoneum. Despite aggressive resuscitative measures, the infant expires within 24 h. Focal hemorrhage, necrosis, and ulceration of which location is most likely be seen at the time of autopsy?

A. Jejunum.
B. Duodenum.
C. Terminal ileum and cecum.
D. Sigmoid colon.

Answer to Question 2:

(C) NEC may involve one or more segments of the GI tract. However, it is most frequently seen in the terminal ileum, with around 40% of cases affecting both the small and the large intestine. While involvement of choices A, B, and D may be seen in NEC, involvement of the terminal ileus is most common.

3) An 18-year-old primigravida gives birth at 34 weeks' gestation to a female infant of low birth weight. She has been receiving formula feeds. On the fourth day of life, the infant becomes hypotensive. Examination is significant for abdominal distention, absent bowel sounds, and bloody stool in the diaper. Which of the follow findings on abdominal radiograph suggests the most likely diagnosis?

A. Intramural gas bubbles within the bowel wall.
B. Dilated stomach and proximal duodenum.
C. Diffusely dilated bowel consistent with ileus.
D. Disparity in size of bowel loops and soap-bubbly appearance in the right lower quadrant with absence of small bowel air-fluid levels.

Answer to Question 3:

(A) This infant is presenting with signs and symptoms suggestive of NEC. Choice A describes pneumatosis intestinalis (PI), a pathognomonic finding for NEC and confirms Definite NEC per Bell's modified criteria. PI is seen in 70–80% of cases. PI appears as a train-track lucency within the bowel wall, occurring when gas-producing bacteria penetrate intraluminal disruptions of the bowel wall mucosa.

## Further Reading

Alganabi M, Lee C, Bindi E, Li B, Pierro A. Recent advances in understanding necrotizing enterocolitis. F1000 Res. 2019;8:107. https://doi.org/10.12688/f1000research.17228.1.

Alsaied A, Islam N, Thalib L. Global incidence of Necrotizing Enterocolitis: a systematic review and meta-analysis. BMC Pediatr. 2020;20(1) https://doi.org/10.1186/s12887-020-02231-5.

Coppola CP. Necrotizing Enterocolitis. In: Coppola CP, Kennedy AP, Scorpio RJ, editors. Pediatric surgery: diagnosis and treatment. Springer; 2014. p. 123–6.

Ein SH, Marshall DG, Girvan D. Peritoneal drainage under local anesthesia for perforations from necrotizing enterocolitis. J Pediatr Surg. 1977 Dec;12(6):963–7. https://doi.org/10.1016/0022-3468(77)90607-8.

Ein SH, Shandling B, Wesson D, Filler RM. A 13-year experience with peritoneal drainage under local anesthesia for necrotizing enterocolitis perforation. J Pediatric Surg. 1990;25(10):1034–7. https://doi.org/10.1016/0022-3468(90)90213-s.

Gilchrist BF. Necrotizing Enterocolitis. first ed. Boca Raton, FL: CRC Press; 2018.

Glynn LA. Necrotizing Enterocolitis. In: Saclarides TJ, Myers JA, Millikan KW, editors. Common surgical diseases an algorithmic approach to problem solving. third ed. Springer; 2015. p. 299–301. chap 76.

Hall NJ. Necrotizing Enterocolitis. In: Davenport M, Geiger JD, editors. Operative pediatric surgery. eighth ed. CRC Press; 2021. p. 368–78. chap 42.

Holcomb GW, Murphy JP, St Peter SD. Chapter 33 on necrotizing enterocolitis. In: Holcomb and Ashcraft's pediatric surgery seventh ed. Elsevier; 2020:536–556.

Jones IH, Hall NJ. Contemporary outcomes for infants with necrotizing enterocolitis—a systematic review. J Pediatr. 2020;220:86–92.e3. https://doi.org/10.1016/j.jpeds.2019.11.011.

Lessin MS, Luks FI, Wesselhoeft CW Jr, Gilchrist BF, Iannitti D, DeLuca FG. Peritoneal drainage as definitive treatment for intestinal perforation in infants with extremely low birth weight (<750 g). J Pediatr Surg. 1998 Feb;33(2):370–2. https://doi.org/10.1016/s0022-3468(98)90465-1.

Mishra P, Foley D, Purdie G, Pringle KC. Intestinal perforation in premature neonates: the need for subsequent laparotomy after placement of peritoneal drains. J Paediatr Child Health. 2015;52(3):272–7. https://doi.org/10.1111/jpc.13013.

Neu J, Walker WA. Necrotizing enterocolitis. N Engl J Med. 2011;364(3):255–64. https://doi.org/10.1056/nejmra1005408.

Niño DF, Sodhi CP, Hackam DJ. Necrotizing enterocolitis: new insights into pathogenesis and mechanisms. Nat Rev Gastroenterol Hepatol. 2016;13(10):590–600. https://doi.org/10.1038/nrgastro.2016.119.

Pierro A, Hall N. Surgical treatment of infants with necrotizing enterocolitis. Semin Neonatol. 2003;8(3):223–32. https://doi.org/10.1016/S1084-2756(03)00025-3.

Quiroz HJ, Rao K, Brady A-C, Hogan AR, Thorson CM, Perez EA, et al. Protocol-driven surgical care of necrotizing enterocolitis and spontaneous intestinal perforation. J Surg Res. 2020;255:396–404. https://doi.org/10.1016/j.jss.2020.05.079.

Sylvester KG, Liu GY, Albanese CT. Necrotizing enterocolitis. In: Coran AG, editor. Pediatric surgery. seventh ed. Elsevier Saunders; 2012. p. 1187–207. chap 94.

Tepas JJ III, Sharma R, Hudak ML, Garrison RD, Pieper P. Coming full circle: an evidence-based definition of the timing and type of surgical management of very low-birth-weight (o1000g) infants with signs of acute intestinal perforation. J Pediatr Surg. 2006;41:418–22.

# Congenital Diaphragmatic Hernia

**Sean C. Skinner**

**Key Points**

1) Defect in the development of the diaphragm.
2) Main concern is the pulmonary hypertension and hypoplasia which affects patient outcome and survival.
3) Repair is not emergent and is institutional dependent.

## Congenital Diaphragmatic Hernia

This is a condition that is characterized by a defect in the diaphragm, allowing intestinal contents to travel into the thoracic cavity and interfering with normal development of the lung. This leads to pulmonary hypoplasia and pulmonary hypertension. Through advanced neonatal intensive care and select application of extracorporeal membrane oxygenation, children can survive to correction.

1. Epidemiology
   a. 0.8–5 per 10,000 births
   b. May be higher in stillborns (1/3 of CDH)
   c. Higher in males
   d. Two percent in first-degree relative
   e. Eighty percent left-sided
   f. Linked to phenmetrazine, thalidomide, quinine, nitrofen, vitamin A deficiency

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2. Associated anomalies are present in 28–50% of infants with CDH and in 100% of stillborn infants with CDH. Common defects are neural tube, cardiac, and midline defects, such as Pentalogy of Cantrell. CDH is more common in Trisomy 13, 18, and 21.
3. Pathophysiology
   a. Diaphragm develops at 4 weeks of gestation.
   b. Midgut returns to abdomen week 9–10.
   c. Defect thought to be caused by impaired development of the post hepatic mesenchymal plate.
   d. If diaphragm open, herniates to thoracic cavity.
   e. Occurs by week 12.
   f. Malrotation occurs.
   g. Hernia sac present in 10–15%.
   h. Left more common than right.
   i. Bochdalek (posterior lateral) hernia is more common (70–75%) than Morgagni (anterior) hernia (13%) (Fig. 1).
   j. Can contain liver, spleen, GI tract, and kidney.
   k. Associated pulmonary hypoplasia
      i. Total pulmonary vascular bed reduced
      ii. Decreased bronchial branching
      iii. Decreased alveoli
      iv. Decreased total vascular diameter and number of vessels
      v. Alveolar vessels remodel and develop medial hyperplasia
      vi. Smooth muscle in alveolar vessels
      vii. Bilateral lungs affected

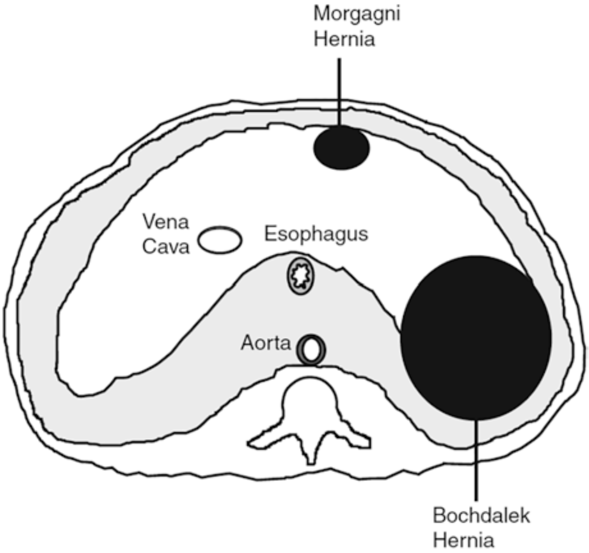

**Fig. 1** Types of congenital diaphragm hernias. Morgagni hernias are defects in the anterior diaphragm, while the more common Bochdalek hernias are in the posterolateral diaphragm. (Source: Christopher Coppola)

l. Factors resulting in persistent fetal circulation in CDH
    i. Pulmonary hypoplasia
    ii. High pulmonary vascular resistance
    iii. Hypoxia
    iv. Acidosis
    v. Stress

m. Diagnosis
    i. Prenatal U/S (CDH, polyhydramnios)
    ii. Fetal MRI—calculate lung-to-head ratio (LHR) and observed to expected LHR
    iii. Respiratory distress after delivery
    iv. Scaphoid abdomen/funnel chest
    v. X-ray with NGT in chest
    vi. Ten to twenty percent with delayed presentation

n. Predicting severity of CDH
    i. Prenatal diagnosed CDH as a group has worse prognosis than CDH diagnosed after birth.
    ii. Presence of cardiac defects worsens mortality.
    iii. Smaller pulmonary artery size (which can be quantified by the McGoon Index) carries worse prognosis.
    iv. Defects large enough to allow herniation of the stomach/liver into the chest have worse prognosis.
    v. Right-sided hernia represents more severe disease occurring earlier in gestation.
    vi. Prenatal LHR calculated:
        1. LHR $< 0.6$ carries poor prognosis.
        2. LHR 0.6–1.0 is a gray zone and careful management.
        3. LHR $> 1.35$ suggests survival.

o. Prenatal care
    i. When a patient is prenatally diagnosed with CDH, parents should receive genetic counseling, and karyotyping should be performed on the fetus to detect any associated anomalies or syndromes.
    ii. Prenatal care should be provided to allow delivery to occur as close as possible to term to minimize the effect of premature delivery on an already compromised pulmonary function.
    iii. When possible, delivery should occur in a center with access to high-level NICU care and ability to perform ECMO, if it becomes necessary.

p. Treatment
    i. Initial resuscitation

1. Patient should be assessed by neonatologist and surgeon after delivery.
2. Consider intubation immediately after delivery to prevent air filling the GI tract and impairing lung function further.
3. IV fluid should be started and used judiciously, 20 ml/kg as a goal to begin, if unresponsive, consider vasopressors early to prevent pulmonary edema.
4. Nasogastric tube placed to decompress the stomach, which may be in the chest, and to reduce the volume of gas and fluid in the GI tract.
5. Pulmonary function should be monitored for signs of pulmonary hypoplasia and failure. The patient may worsen over the course of the initial 24–48 h "honeymoon" period.
6. Obtain chest X-ray to detect abdominal organs in the chest and degree of hypoplasia.
7. When intubation and mechanical ventilation is needed, use a strategy of "gentle ventilation" to avoid barotrauma to the alveoli.
    a. Conventional ventilation with peak pressures less than 25 cm $H_2O$.
    b. High-frequency/low-pressure ventilation, with use of an oscillating/jet ventilator when needed.
    c. Goal of preductal saturation 85–95%, post-ductal saturations >70%.
    d. Permissive hypercapnia with $PCO_2$ 45–60 mmHg.
    e. Medications to reduce pulmonary hypertension.
        i. Inhaled nitric oxide
        ii. Sildenafil
        iii. Bosentan (Tracleer)
        iv. Epoprostenol (Flolan)
        v. Tolazoline
        vi. Inhaled prostacyclin
    f. When gentle ventilation and all adjuncts fail to provide adequate oxygenation, evaluate patient for suitability of ECMO therapy.

ii. Surgical repair of CDH
    1. Timing of operation: multiple options to timing.
        a. In stable and uncomplicated case—Repair when weaned on $FiO_2$ and minimal ventilator settings.
        b. In complicated case—requiring ECMO.
            i. "Early" repair—Once stable on ECMO, repair within first 24–48 h before develops edema and risk of bleeding increases.
            ii. "Delayed" repair—While on ECMO and beginning to wean, once close to coming off ECMO, repair prior to decannulation in case develops pulmonary hypertensive crisis from stress of surgery.
            iii. "Late" repair—Recovered and off ECMO, then repair.

2. Technique
    a. Subcostal incision or thoracotomy: On the left, the approach via abdomen allows for convenient reduction of viscera into abdominal cavity. For a right-sided CDH, thoracotomy avoids the difficulty of working around the liver that is encountered with an abdominal approach.
        i. Reduce hernia contents back into abdomen.
        ii. Excise hernia sac when present as it can interfere with visualization of lungs or viscera while closing defect.
        iii. Identify muscular rim of diaphragm circumferentially. Some areas along posterolateral ribs and mediastinal structures may have no diaphragm and require careful placement of sutures.
        iv. Determine if primary closure is possible or if a patch of exogenous material (artificial or biologic mesh) or muscle flap will be needed (approximately one-third of cases).
        v. Consider addressing malrotation/non-fixation of bowel, if present, by performing a Ladd's procedure.
        vi. Close abdomen if possible. If there is insufficient domain in abdomen after reducing viscera from chest to abdomen, a silo or mesh closure of abdomen may be needed on a temporary basis.

q. Outcome
    i. Through the twentieth century, CDH has historically had 50% survival; however advancements in neonatal intensive care unit techniques and select use of ECMO have produced a survival range of 39–95% (mean, 69%), depending on setting of care.
    ii. There is a risk of both early and late recurrence. Early recurrence can be due to technical error and increased abdominal pressure. Placement of mesh reduces tension across the repair, but approximately one-third of patients will need a late revision of repair due to recurrence of hernia or due to tension on the thoracic wall and deformation of the ribcage.
    iii. Chronic lung disease may result from the accompanying pulmonary hypoplasia.
    iv. Gastroesophageal reflux is common after CDH repair, and some patients will require an anti-reflux procedure.
    v. Development/cognitive deficits occur and can be due to associated anomalies or as sequela from ECMO.

## Additional Notes

Congenital diaphragmatic hernia is a life-threatening condition of the newborn in which a gap in the diaphragm allows abdominal contents to enter the chest. There are varying degrees of severity, and often the accompanying pulmonary hypoplasia and pulmonary hypertension are the main determinants of long-term outcome. Historically, there has been a 50% mortality rate, but the recent reports show a survival rate that approaches 90% [1]. High-volume centers treating larger numbers of patients achieve the highest survival and will have greater resources for more intense therapy like ECMO. The majority of advances have been in neonatal intensive care and pharmaceutical therapy, but some operative alternatives are thoracoscopic approach and creation of a dome shape to the repair [2, 3]. Elevated intraabdominal pressure measured via the bladder after repair is predictive of length of mechanical ventilation and delay before enteral feeding [4]. Duration of mechanical ventilation and duration of nitric oxide treatment predict the 11% of survivors who will require pulmonary hypertension medication after discharge [5].

## Study Questions

1. Question 1: What is the most common type of CDH?
    a. Morgagni
    b. Bilateral
    c. Bochdalek
    d. Anterior

Answer: (c.) Bochdalek. The most common type of CDH is Bochdalek (80%), the most common type of defect is posterior lateral. Anterior defects or Morgagni are more likely delayed in presentation and less common. Bilateral CDH are rare and usually fatal.

2. Question 2: What are the goals of initial management in a newborn with CDH?
    a. Bag ventilation, monitor to see how baby does, feed when ready.
    b. Immediate intubation, gastric decompression, gentle ventilation.
    c. Bag ventilation, fluid resuscitation as need, up to 60 ml/kg.
    d. Gastric decompression and high-frequency ventilation with high pressures.

Answer: (b.) Immediate intubation, gastric decompression, gentle ventilation. Goal in CDH is to intubate early to avoid GI filling with air, gastric decompression, gentle ventilation with low-pressure ventilation and high frequency. Do not want high-pressure ventilation to prevent barotrauma in already compromised lungs.

3. Question 3: What can predict poor outcome or difficult management CDH?
    a. LHR = 1.0, spleen in chest, $SpO_2 = 90\%$.
    b. LHR 1.35, stomach in abdomen, $PCO_2 = 40$.
    c. LHR = 1.0, stomach in chest, peak vent pressure 20 mmHg.
    d. LHR <0.6, liver in chest, stomach in chest, $PCO_2 > 70$.

Answer: (d.) LHR < 0.6, liver in chest, stomach in chest, $PCO_2 > 70$. Lung-to-head ratio <0.6 portends a bad prognosis, liver and stomach in the chest can be risk for poor outcome, high $PCO_2$ can suggest pulmonary atresia is more severe and poor prognosis.

## References

1. Bose S, Apeksha D, White B, Laje P, Hedrick H, Peranteau WH. National congenital diaphragmatic hernia outcomes: assessing volume-outcomes relationships using the 2016 KID database. J Am Coll Surg, Pediatr Surg. 2021;233(5 Supp. 1):S180.
2. Kotb M, Shehata S, Khairi A, Mohamed Shehata S, Ghoneim T, Rabie A. Thoracoscopic repair of neonatal congenital diaphragmatic hernia: Minimizing Open repair in a low-income country. J Laparoendosc Adv Surg Tech A. 2021;31(11):1341–45.
3. Verla MA, Style CC, Lee TC, Menchaca AD, Lau PE, Mehollin-Ray AR, Fernandes CJ, Keswani SG, Olutoye OO. Does creating a dome reduce recurrence in congenital diaphragmatic hernia following patch repair? J Pediatr Surg. 2022;57(4):637–42.
4. Thambusamy E, Thambidorai CR, Nah SA, Gan CS, Wang J, Singaravel S, Sanmugam A. Intra-abdominal hypertension in neonates following congenital diaphragmatic hernia repair: Correlation with early postoperative respiratory and gastrointestinal outcomes. J Pediatr Surg. 2022;57(2):199–202.
5. Mahmood B, Murthy K, Rintoul N, Weems M, Keene S, Brozanski B, DiGeronimo R, Haberman B, Hedrick H, Gien J, Seabrook R, Ali N, Chapman R, Daniel J, Harrison A, Johnson Y, Porta NFM, Uhing M, Zaniletti I, Grover TR; Children's Hospitals Neonatal Consortium. Predicting treatment of pulmonary hypertension at discharge in infants with congenital diaphragmatic hernia. J Perinatol. 2022;42(1):45–52.
6. Chandrasekharan, et al. Congenital diaphragmatic hernia – a review. Maternal Health, Neonatology, and Perinatology. 2017;3(6)
7. Lally KP. Congenital diaphragmatic hernia – the past 25 (or so) years. J Pediatr Surg. 2016;51:695–8.
8. McHoney M. Congenital diaphragmatic hernia, management in the newborn. Pediatric Surg Int. 2015;32:1005–13.
9. Hollinger LE, et al. Long-term follow-up of congenital diaphragmatic hernia. Semin Pediatr Surg. 2017;26:178–84.
10. Puligandla PS, et al. Management of congenital diaphragmatic hernia: a systematic review from the APSA outcomes and evidence based practice committee. J Pediatr Surg. 2015;50:1958–70.
11. Snoek KG, et al. Standardized postnatal management of infants with congenital diaphragmatic hernia in Europe: The CDH EURO Consortium Consensus – 2015 Update. Neonatology. 2016;110:66–74.
12. Yang MJ, et al. Congenital diaphragmatic hernia: a narrative review of controversies in neonatal management. Transl Pediatr. 2021;10(5):1432–47.
13. Morini F, et al. Ventilation modalities in infants with congenital diaphragmatic hernia. Seminars in Pediatric Surgery. 2017;26(3):159–65.

# Esophageal Atresia and Tracheoesophageal Fistula

**Robert Ricca and Kristine Griffin**

**Key Points**

1. Esophageal atresia/tracheoesophageal fistula (EA/TEF) is an interruption of the normal formation of the esophagus. The most common variant seen is an esophageal atresia with a tracheoesophageal fistula to the distal esophagus.
2. All children diagnosed with EA/TEF require a VACTERL (vertebral, anorectal, cardiac, tracheoesophageal, renal, limb) workup for associated anomalies to include a physical examination, echocardiogram, renal ultrasound, and plain radiographs of the spine. The echocardiogram should be performed prior to operative intervention.
3. Surgical correction can be performed through an open thoracotomy or thoracoscopically based upon the surgeon's experience and comfort.
4. Many children with EA/TEF will develop gastroesophageal reflux disease, tracheomalacia, or an esophageal stricture at the anastomosis which may prompt further operative treatments.
5. Long-gap esophageal atresia is best treated by a delayed operative intervention to allow for growth of the esophageal pouch. Multiple options are available for reconstruction or replacement of the native esophagus and depends upon institutional experience.

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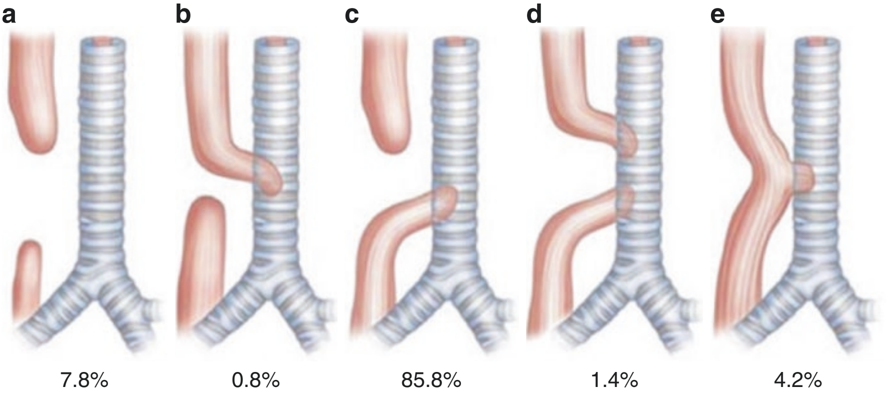

**Fig. 1** Gross classification of esophageal atresia and tracheoesophageal fistula. (**a**) Pure esophageal atresia without fistula; (**b**) proximal tracheoesophageal fistula, blind distal pouch; (**c**) distal tracheoesophageal fistula, blind proximal pouch; (**d**) proximal and distal tracheoesophageal fistula; (**e**) intact continuous esophagus with H-type tracheoesophageal fistula. Numbers below each type are percentage of children with esophageal atresia/tracheoesophageal fistula presenting with that type. (Source: Bryan Walters)

Esophageal atresia is an interruption of the normal formation of the esophagus. Classification of esophageal atresia is based on the presence or absence of a fistula to the trachea.

1. Pathophysiology:
    a. Types of esophageal atresia/tracheoesophageal fistula (Gross classification): (Fig. 1)
        i. Type A: Esophageal atresia with no tracheoesophageal fistula, 7.8% (pure esophageal atresia).
        ii. Type B: Esophageal atresia and a proximal tracheoesophageal fistula, 0.8%.
        iii. Type C: Esophageal atresia and a distal tracheoesophageal fistula, 85.8% (most common type).
        iv. Type D: Esophageal atresia with both a proximal and distal tracheoesophageal fistula, 1.4%.
        v. Type E: Fistula to the trachea with no esophageal atresia, 4.2% (H-type fistula).
    b. Embryology:
        i. The esophagus and trachea arise from the fetal foregut which appears at 3 weeks gestation and develop into separate structures up to the level of the larynx by the fourth week of gestation. The defect occurs by the 20th week of gestation with rapid growth of the trachea. There is continued debate regarding the embryopathogenesis of esophageal atresia.

- c. Incidence: 1 in 4000–5000 live births.
  - i. According to a recent study:
    1. About 13% were diagnosed prenatally.
    2. Median gestational age at birth is 37 weeks.
    3. Median birth weight is 2.6 kg.
    4. Gender ratio M/F is 1.55 [1].
- d. Associated anomalies:
  - i. Maternal polyhydramnios: 60% of patients.
  - ii. Cardiac: 30% with patent ductus arteriosum, ventricular septal defect, or atrial septal defect.
  - iii. Gastrointestinal: 12% have imperforate anus or duodenal atresia.
  - iv. VACTERL constellation of anomalies: 20–30% of cases.
    1. V: Vertebral
    2. A: Anorectal
    3. C: Cardiac
    4. TE: Tracheoesophageal
    5. R: Renal
    6. L: Limb (e.g. absent or hypoplastic radial bone)

2. Clinical presentation:
   - a. Prenatal findings: polyhydramnios with or without small gastric bubble (seen more often with pure esophageal atresia).
   - b. Symptoms: excessive drooling, choking, coughing, and episodes of cyanosis, especially with attempts to feed.
3. Diagnosis:
   - a. Inability to pass a nasogastric tube.
     - i. NG tube may coil above level of esophageal atresia.
     - ii. For confirmation, a small amount of barium contrast may be injected into the NG tube under fluoroscopic guidance. Gastrograffin should be avoided due to risk of pneumonitis.
     - iii. Children with an H-type fistula or Type E EA/TEF may have a delayed presentation due to the absence of an atretic esophagus. These children may have frequent respiratory events during feeding and can be diagnosed with a prone video esophagram or bronchoscopy. A high index of suspicion for an H-type fistula is necessary in children with recurrent aspiration events.
4. Initial management:
   - a. Prevent aspiration: keep patient NPO, keep head up, place NG or OG tube in proximal pouch, and maintain suction.

- b. Rule out associated abnormalities: examination of anus and limbs, ultrasound of kidneys, cardiac echocardiogram, plain film of the vertebrae, and consideration of spinal ultrasound if vertebral anomalies present.
- c. Echocardiogram must be performed preoperatively to evaluate for congenital cardiac conditions as well as to ensure a left-sided aortic arch.
- d. Preferable to avoid positive pressure ventilation to prevent gastric distention.

5. Treatment:
    - a. Preoperative antibiotics
    - b. Intubate patient in OR (consider bronchoscopic airway evaluation at this time to evaluate for presence of multiple fistulae)
    - c. Surgical correction:
        - i. Open approach:
            1. Right lateral thoracotomy
            2. Perform extrapleural dissection (if possible)
            3. Divide azygous vein
            4. Divide tracheoesophageal fistula if present
            5. Close trachea
            6. Anastomose proximal and distal ends of esophagus
            7. Place right retropleural chest tube
            8. Close thoracotomy
        - ii. Thoracoscopic approach:
            1. Right thoracoscopy, typically in left lateral decubitus position or prone with right side slightly up.
            2. Camera port and two working ports placed.
            3. Chest is insufflated with maximum pressure of 3–5 mmHg and flow of 1 L/min.
            4. Azygous vein may be divided or preserved.
            5. Divide fistula.
            6. Close trachea.
            7. Anastomose proximal and distal ends of esophagus.
            8. Place chest tube.
            9. Close ports [2].
        - iii. Use of a transanastomotic feeding tube (TAFT) has been advocated by some surgeons to allow for early enteral feeding and to stent open the esophageal anastomosis. This practice remains controversial as other studies have shown an increased rate of esophageal stricture in children who have a TAFT placed [3].
        - iv. Currently, most EA/TEF repairs are being done via open approach. Thoracoscopic approach is gaining popularity at high-volume centers. Operative time tends to be longer with the minimally invasive approach.

      v. A recent meta-analysis showed thoracoscopic repair had shorter length of hospital stay and shorter interval to first oral feed. There were no differences in leak rate, strictures, pulmonary complications, rates of fundoplication, or blood loss [4].

   d. Postoperative care:

      i. Early extubation, adequate pain control, early enteric feeds via transanastomotic gastric tube, if placed, and placement of chest tube to water seal if no air leak.
      ii. One week after operation, obtain contrast esophagram via oral feeding of contrast with bottle or through previously placed nasogastric tube. If no leak present, the nasogastric tube and the chest tube can be removed. If a leak is present, maintain the tubes and repeat the study in 1 week.

6. Complications:

   a. Anastomotic leak: occurs in approximately 20% of patients.

      i. If leak is evident on contrast study, can continue to feed via transanastomotic tube. If tube has been removed, make patient NPO. Contrast study should be repeated in 1 week to monitor for resolution of leak.
      ii. Glycopoyrrolate (Robinul) has been used as an adjunct in patients with leaks. By decreasing salivary secretions, it may promote spontaneous closure of anastomotic leak [5].

   b. Gastroesophageal reflux disease: approximately 13% of patients who undergo repair of a tracheoesophageal fistula repair will require an antireflux procedure, of which the Nissen fundoplication is most commonly performed (Lal et al. ).
   c. Tracheomalacia: evidenced by a barking cough present for up to 1 year after surgical correction of the tracheoesophageal fistula. Severe cases may require aortopexy.

      i. About 2% of patients will require aortopexy [1].
      ii. Some surgeons perform prophylactic posterior tracheopexy at the time of primary esophageal atresia repair for patients with a flaccid posterior tracheal membrane. This is done by placing several non-absorbable sutures between the posterior tracheal wall to the anterior longitudinal spinal ligament [6].

   d. Stricture: occurs in 30% of repairs. Treat with dilation of esophagus using a radial expanding balloon dilators in the operating room. This can be done in a sequential fashion using fluoroscopic guidance. Early identification of the stricture and early intervention with dilation increase the likelihood of success. Injection of triamcinolone may aid resolution.
   e. Recurrent fistula: treat with fibin glue plug, cauterization of fistula, or repeat suture if fails to close with time and conservative measures.

7. Treatment strategy for pure esophageal atresia without fistula [7, 8]:
    a. Diagnosis is the same as esophageal atresia with tracheoesophageal fistula; however, abdominal X-ray will show a gasless abdomen. Esophageal atresia with a proximal fistula may show similar findings. Bronchoscopy can confirm the presence or absence of a proximal fistula.
    b. Treatment:
        i. Maintain suction catheter in upper pouch and operative creation of gastrostomy. Obtain "gapogram": orogastric tube in upper pouch and dilator in lower pouch via gastrostomy. If gap between upper pouch and lower pouch is less than three vertebrae, then child is a candidate for repair of esophagus by primary anastomosis. If distance between pouches is greater, the patient has long-gap esophageal atresia; the optimum treatment is controversial.
        ii. Delayed repair is typically considered the initial option for management of long-gap esophageal atresia to allow for growth of the proximal pouch. The distal esophagus may also be elongated by attaching it to the spine to ensure elongation during this growth phase. A period of time varying from 6 weeks to 9 months may be considered.
        iii. Foker technique: tension-induced lengthening of the esophagus through traction repair may be used in long-gap esophageal atresia but is controversial. Traction sutures are placed on both ends of the esophagus and are secured internally to prevertebral fascia or brought out through the chest wall. The esophageal segments are wrapped in silastic sheeting to decrease adhesions. Postoperatively, for external traction, tension is gradually increased on a daily basis by placing pieces of feeding tubes under the sutures. Tension is maintained on these sutures to elongate the esophagus until sufficient lengthening has occurred so that primary anastomosis is possible. Clips are placed on each esophageal segment intraoperatively and progress is monitored by serial X-rays. Weekly contrast studies are performed to confirm lengthening and to observe potential esophageal leaks. The esophageal pouches are elongated in this manner until they can tolerate a primary anastomosis. This may require several operations.
        iv. Other options for increased length of the esophageal pouches include circular or spiral myotomy of the proximal pouch due to the robust blood supply. Increased length of the distal pouch has been reported through the use of a partial gastric mobilization or creation of a gastric tube from the proximal stomach.
        v. If esophageal lengthening procedures fail, the esophagus can be replaced with gastric pull-up, reverse gastric tube, small bowel, or colon. Studies to date have not shown one approach to be beneficial over another but is based upon institutional experience.

## Laryngeal Tracheal Cleft

Laryngeal, laryngotracheal, or laryngotracheoesophageal clefts (henceforth referred to as laryngeal clefts) are a spectrum of rare anomalies which allow communication between the trachea and esophagus. They occur in 1 in 10,000–20,000 live births, with a male predominance [9].

1. Pathophysiology
   a. Laryngeal clefts result from a failure of fusion of the tracheoesophageal septum or of lateral growth of the posterior cricoid cartilage [10].
2. Associated anomalies
   a. Syndromes associated with laryngeal clefts include Opitz Frias syndrome (craniofacial anomalies, cleft lip and palate, hypertelorism), Pallister Hall syndrome (polydactyly, imperforate anus, hypothalamic/pituitary, renal, cardiac, pulmonary abnormalities), VACTERL, and CHARGE syndrome (coloboma, heart anomalies, choanal atresia, growth retardation, genital anomalies, ear anomalies) [11].
3. Presentation
   a. Patients with a posterior laryngeal cleft typically present with chronic cough, especially with feeds, and respiratory distress. Symptoms are usually specific to type of cleft.
   b. Benjamin and Inglis classification and presenting symptoms [11, 12] (Fig. 2):
      i. Type I: supraglottic interarytenoid cleft is above the level of vocal cord
         - Stridor, toneless or hoarse cry, hypersecretions, swallowing disorders, cyanosis during feeding
      ii. Type II: cleft extends below vocal cords into upper cricoid cartilage
         - Aspiration and recurrent pneumonia
      iii. Type III: cleft extends through cricoid cartilage and possibly into cervical trachea
         - Aspiration and recurrent pneumonia
      iv. Type IV: cleft extends into thoracic trachea and extends toward the carina
         - Early respiratory distress with bronchial flooding, difficulty maintaining adequate ventilation

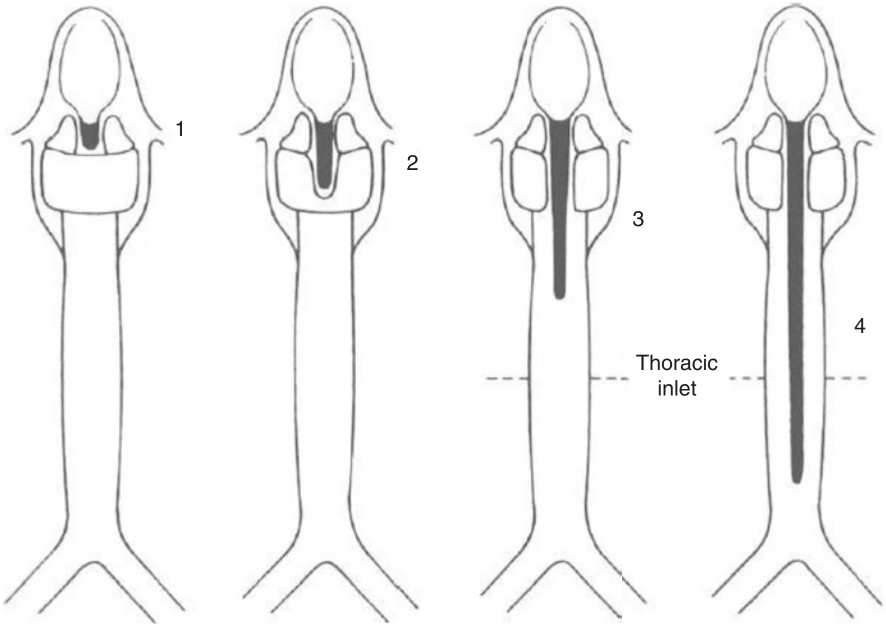

**Fig. 2** Benjamin–Inglis classification of laryngotracheal cleft. Type I: supraglottic interarytenoid, cleft limited to at or above true vocal cords. Type II: cleft involves level of true vocal folds, partial involvement of cricoid lamina. Type III: cleft extends completely through cricoid. Type IV: cleft extension includes posterior wall of thoracic trachea below cricoid. (Source: Leboulanger N, Garabédian EN. Laryngo-tracheo-oesophageal clefts. Orphanet J Rare Dis 6:81, 2011. https://doi.org/10.1186/1750-1172-6-81. Used under Creative Commons Attribution License)

4. Diagnosis
    a. Lateral chest X-rays may show abnormal communication and lack of soft tissue between trachea and esophagus. Anteroposterior chest X-rays may only show evidence of aspiration manifested as pulmonary parenchymal abnormalities [13].
    b. Barium swallow study in search of a tracheoesophageal fistula may show immediate passage of barium into the trachea. This could lead to a false diagnosis of a tracheoesophageal fistula [14].
    c. Laryngoscopy is used for definitive diagnosis. Bronchoscopic examination should be conducted to evaluate for associated tracheobronchial abnormalities [11].

5. Medical management
    a. Initial medical management includes antireflux medication, feeding precautions, and optimization of respiratory status [15].

6. Surgical management
    a. Endoscopic repair
        i. Type I and II, and select Type III clefts: Interarytenoid mucosa is incised from the arytenoid on one side down through the bottom of the cleft and up to the arytenoid on the other side. Mucosal flaps are made on either side and double layer closure is performed [16].
    b. Open repair
        i. Anterior translaryngotracheal approach: a vertical incision is made in the thyroid cartilage, cricoid cartilage, and first two tracheal rings. The laryngeal cleft is exposed, and mucosa is incised. The cleft is closed. The posterior pharynx is closed and interposition graft (using tibial periosteum, muscle flap, pleura, etc.) placed over repair. The anterior larynx is closed. The airway is maintained through a separate tracheotomy throughout the procedure.
        ii. Special cases:
            1. Partial upper sternotomy with cervical incision: performed for Type IV clefts.
            2. Anterior cervico-thoracic approach: requires ECMO [11].

## CHAOS

Congenital high airway obstruction syndrome, or CHAOS, is complete or partial obstruction of the fetal airway. If unrecognized prenatally, it may lead to stillbirth or postnatal death [17].

1. Pathophysiology
    a. CHAOS is caused by failure of the upper respiratory tract to recanalize around 10 weeks of gestation [18]. This causes obstruction of fetal lung fluid, leading to hyperexpansion of the lungs. The resultant increased intrathoracic pressure diminishes venous return and cardiac output [19].
2. Presentation
    a. CHAOS may present as a complete laryngeal atresia without esophageal fistula, complete laryngeal atresia with TEF, or a near-complete high upper airway obstruction [20].
3. Diagnosis
    a. Prenatal ultrasound findings of bilateral large lungs, dilated tracheobronchial tree, compressed heart, flattened diaphragm, and ascites suggest CHAOS [21].

4. Management

   a. Fetuses with suspected CHAOS syndrome should be delivered using the *ex utero* intrapartum treatment, or EXIT, procedure. This allows time to establish a potentially difficult neonatal airway in a controlled manner by delaying cessation of placental circulation. A multidisciplinary team including obstetricians, neonatologists, anesthesiologists, and otolaryngologists is necessary to complete the delivery. The mother is placed under general anesthesia to facilitate uterine relaxation and uteroplacental gas exchange. A cesarean section is performed. The fetal head and thorax are delivered and pulse oximeter is placed. The fetal airway is then secured using direct laryngoscopy, bronchoscopy, or other advanced airway techniques. Tracheostomy can be performed if initial attempts at intubation are unsuccessful. Once the airway is secured, the delivery is completed and umbilical cord is clamped [22]. Once the child has stabilized and grown, the airway can be reconstructed by an otolaryngologist [20].

## Additional Notes

Esophageal atresia was first described by Durston in 1670, although Gibson in the 1700s made the most accurate description with his necropsy findings of a child with esophageal atresia and distal trachea-esophageal fistula. Dr. Thomas Landman made five unsuccessful attempts at primary repair. Cameron Haight performed the first successful primary repair in 1941 after three failed procedures. Drs. Leven and Ladd had the first long-term survivors using serial operations. The last several decades have shown a focus on the best operative management for children with long-gap esophageal atresia in an effort to save the native esophagus. Lengthening procedures using traction-induced growth as well as replacement with gastric pull-up, colonic replacement, or jejunal replacement have all been described in the literature [8, 23].

## Study Questions

1) A newborn infant undergoes a repair of an esophageal atresia with distal trachea-esophageal fistula. He is found to have severe tracheomalacia on dynamic bronchoscopy at the beginning of the operative procedure. What procedure should the surgeon consider at the time of EA/TEF repair?

   a. Aortopexy
   b. Posterior tracheopexy
   c. Delay surgical repair of the atresia
   d. Tracheostomy

Answer: (b) In this child, it would be reasonable to consider a posterior tracheopexy at the time of operative repair. There would be no indication for delayed repair or a tracheostomy. While aortopexy has been described for the treatment of tracheomalacia, recent literature has shown the efficacy of a posterior tracheopexy at the time of surgical repair of the esophageal atresia.

2) A newborn infant is found to have esophageal atresia with no fistula present and a gasless abdomen. The proximal pouch ends at the thoracic inlet. What is the best initial management for this child with pure gap esophageal atresia.

    a. Gastrostomy tube, confirm long gap and delayed operative repair
    b. Immediate placement of traction-induced growth sutures (Foker)
    c. Colonic interposition graft
    d. Primary repair of esophagus

Answer: (a) This child presents with a long-gap esophageal atresia. The best initial management based upon the current literature would be a delayed operative approach with placement of a gastrostomy tube. This can allow for continued enteral feeds with growth of the proximal and distal pouch. Some surgeons advocate tacking the distal pouch to the spine to assist with growth of the distal segment.

3) A 2-month-old infant presents with a history of recurrent aspiration events with fistula. He is found to have an H-type or Type E tracheoesophageal fistula with no atretic segment. The best operative approach for this patient is through:

    a. Right neck incision
    b. Right thoracotomy
    c. Left neck incision
    d. Left thoracotomy

Answer: (a) The H-type tracheoesophageal fistula or TEF with no esophageal atresia is best approached through a right neck incision. These fistulae tend to be higher and can be best reached through a neck incision. Placement of a Fogarty catheter or other catheter through the fistula on initial bronchoscopic examination can assist with identification of the fistula in the operating room.

## References

1. Lal DR, Gadepalli SK, Downard CD, Ostlie DJ, Minneci PC, et al. Perioperative management and outcomes of esophageal atresia and tracheoesophageal fistula. J Pediatr Surg. 2017;52(8):1245–51.
2. Rozeik AE, Elbarbary MM, Saleh AM, Khodary AR, Al-Ekrashy MA. Thoracoscopic versus conventional open repair of tracheoesophageal fistula in neonates: a short-term comparative study. J Pediatr Surg. 2020;55(9):1854–9.
3. Wang C, Feng L, Li Y, Ji Y. What is the impact of the use of transanastomotic feeding tube on patients with esophageal atresia: a systematic review and meta-analysis. BMC Pediatr. 2018;18(1):385.

4. Wu Y, Kuang H, Lv T, Wu C. Comparison of clinical outcomes between open and thoracoscopic repair for esophageal atresia with tracheoesophageal fistula: a systematic review and meta-analysis. Pediatr Surg Int. 2017;33(11):1147–57.
5. Mathur S, Vasudevan SA, Patterson DM, Hassan SF, Kim ES. Novel use of glycopyrrolate (Robinul) in the treatment of anastomotic leak after repair of esophageal atresia and tracheoesophageal fistula. J Pediatr Surg. 2011;46(3):e29–32.
6. Tytgat SHAJ, van Herwaarden-Lindeboom MYA, van Tuyll van Serooskerken ES, van der Zee DC. Thoracoscopic posterior tracheopexy during primary esophageal atresia repair: a new approach to prevent tracheomalacia complications. J Pediatr Surg. 2018;53(7):1420–3.
7. Bairdain S, Hamilton TE, Smithers CJ, Manfredi M, Ngo P, et al. Foker process for the correction of long gap esophageal atresia: primary treatment versus secondary treatment after prior esophageal surgery. J Pediatr Surg. 2015;50(6):933–7.
8. Baird R, Lal DR, Ricca RL, Diefenbach KA, Downard CD, et al. Management of long gap esophageal atresia: a systematic review and evidence-based guidelines from the APSA outcomes and evidence based practice committee. J Pediatr Surg. 2019;54(4):675–87.
9. Watters K, Ferrari L, Rahbar R. Laryngeal cleft. Adv Otorhinolaryngol. 2012;73:95–100. https://doi.org/10.1159/000334452. Epub 2012 Mar 29
10. Johnston DR, Watters K, Ferrari LR, Rahbar R. Laryngeal cleft: evaluation and management. Int J Pediatr Otorhinolaryngol. 2014;78(6):905–11. https://doi.org/10.1016/j.ijporl.2014.03.015. Epub 2014 Mar 27
11. Pezzettigotta SM, Leboulanger N, Roger G, Denoyelle F, Garabédian EN. Laryngeal cleft. Otolaryngol Clin N Am. 2008;41(5):913–33., ix. https://doi.org/10.1016/j.otc.2008.04.010.
12. Benjamin B, Inglis A. Minor congenital laryngeal clefts: diagnosis and classification. Ann Otol Rhinol Laryngol. 1989;98(6):417–20. https://doi.org/10.1177/000348948909800603.
13. DuBois JJ, Pokorny WJ, Harberg FJ, Smith RJ. Current management of laryngeal and laryngotracheoesophageal clefts. J Pediatr Surg. 1990;25(8):855–60. https://doi.org/10.1016/0022-3468(90)90191-b.
14. Eriksen C, Zwillenberg D, Robinson N. Diagnosis and management of cleft larynx. Literature review and case report. Ann Otol Rhinol Laryngol. 1990;99(9 Pt 1):703–8. https://doi.org/10.1177/000348949009900907.
15. Strychowsky JE, Rahbar R. Laryngotracheoesophageal clefts. Semin Pediatr Surg. 2016;25(3):128–31. https://doi.org/10.1053/j.sempedsurg.2016.02.005. Epub 2016 Feb 21
16. Koltai PJ, Morgan D, Evans JN. Endoscopic repair of supraglottic laryngeal clefts. Arch Otolaryngol Head Neck Surg. 1991;117(3):273–8. https://doi.org/10.1001/archotol.1991.01870150041004.
17. Joshi P, Satija L, George R, Chatterjee S, D'Souza J, Raheem A. Congenital high airway obstruction syndrome-antenatal diagnosis of a rare case of airway obstruction using multimodality imaging. Med J Armed Forces India. 2012;68(1):78–80. https://doi.org/10.1016/S0377-1237(11)60111-1. Epub 2012 Jan 18
18. Hedrick MH, Ferro MM, Filly RA, Flake AW, Harrison MR, Adzick NS. Congenital high airway obstruction syndrome (CHAOS): a potential for perinatal intervention. J Pediatr Surg. 1994 Feb;29(2):271–4. https://doi.org/10.1016/0022-3468(94)90331-x.
19. Lim FY, Crombleholme TM, Hedrick HL, Flake AW, Johnson MP, Howell LJ, Adzick NS. Congenital high airway obstruction syndrome: natural history and management. J Pediatr Surg. 2003 Jun;38(6):940–5. https://doi.org/10.1016/s0022-3468(03)00128-3.
20. Hartnick CJ, Rutter M, Lang F, Willging JP, Cotton RT. Congenital high airway obstruction syndrome and airway reconstruction: an evolving paradigm. Arch Otolaryngol Head Neck Surg. 2002 May;128(5):567–70. https://doi.org/10.1001/archotol.128.5.567.
21. Aslan H, Ekiz A, Acar DK, Aydiner B, Kaya B, Sezer S. Prenatal diagnosis of congenital high airway obstruction syndrome (CHAOS). Five case report. Med Ultrason. 2015;17(1):115–8. https://doi.org/10.11152/mu.2013.2066.171.haek.
22. Prickett K, Javia L. Fetal evaluation and airway management. Clin Perinatol. 2018;45(4):609–28. https://doi.org/10.1016/j.clp.2018.07.003.
23. Nakayama DK. The history of surgery for esophageal atresia. J Pediatr Surg. 2020;55(7):1414–9.

# Extracorporeal Membrane Oxygenation (ECMO)

**Sean C. Skinner**

**Key Points**

1) ECMO is an advanced method to manage reversible cardiorespiratory failure when all medical therapy has been exhausted.
2) There are two modalities of ECMO. Veno-Arterial (VA), which requires two cannulas and blood returns to the systemic circulation. Veno-Venous (VV), which utilizes one cannula and returns oxygenated blood to the venous system.
3) The goal of ECMO is to improve tissue oxygen delivery while waiting for the lungs or heart to recover.

1. Introduction: Extracorporeal Membrane Oxygenation (ECMO) also referred to as Extracorporeal Life Support (ECLS) is a method of cannulating a patient's blood vessels, removing the blood from the patient, oxygenating the blood, removing $CO_2$, warming the blood, and then returning it to the patient. This is performed to support the patient through respiratory or cardiac failure.
2. Indications: Patients who have cardiorespiratory failure who have not responded to maximal therapy including maximum dose of vasopressors, adequate fluid resuscitation, maximum concentration of inhaled nitric oxide (iNO), requiring maximum ventilation strategies including high frequency or high peak pressures as well as 100% oxygen delivery.

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Neonatal/pediatric respiratory or cardiac failure from:
  - Meconium aspiration syndrome (MAS)
  - Persistent pulmonary hypertension (PPHN)
  - Pneumonia (aspiration, bacterial, viral)
  - Air leak syndrome
  - ARDS/asthma/cystic fibrosis
  - Myocarditis
  - Congenital heart disease
  - Congenital diaphragmatic hernia
  - Pulmonary embolus
  - Sepsis
  - Vasopressor and fluid resistant cardio/respiratory failure
  - Bridge to transplantation/donation

3. Criteria: Various criteria are available to help make the decision of when to go on ECMO. Below are a few of the more highly used calculations to help make the decision to place a patient on ECMO.

   a. Oxygenation index (OI):

      i. (Mean airway pressure (MAP) × $FiO_2 \times 100)/PaO_2$.
      ii. $OI > 40$ (80% mortality); $OI = 25–40$ (50% mortality).

   b. Alveolar–arterial gradient (A-a gradient):

      i. $(FiO2\ (713) - PaO_2 - PaCO_2)/PaO_2$.
      ii. A-a > 610 on at least two consecutive blood gases (79% mortality).

   c. P/F ratio:

      i. $PaO_2/FiO_2$ ratio < 100 mm Hg.

   d. Scoring systems:

      i. Pediatric pulmonary rescue with ECMO prediction (P-PREP)—prognostic score to predict mortality at the time of ECMO initiation for children.
      ii. Pediatric risk estimate score for children using extracorporeal respiratory support (Ped-RESCUERS)—designed to estimate the in-hospital mortality risk for children prior to being placed on ECMO.

4. Contraindications:

   a. Gestational age < 32 weeks (increased risk of intraventricular hemorrhage).
   b. Weight less than 2.0 kg (small vessels).
   c. Intracranial hemorrhage (IVH) > Grade II.
   d. Prolonged mechanical ventilation >10–14 days with high mean pressures.
   e. Severe chromosomal anomaly not compatible with life.
   f. Irreversible lung or cardiac pathology.
   g. Prolonged arrest or resuscitation and possible brain death.

5. Types of ECMO:
   a. Veno-Venous (VV).
      i. Placing a double lumen, or two single lumen cannula into a large vein.
      ii. Blood is withdrawn from a large vein and is returned to the large vein and to the right heart.
      iii. Dependent on natural cardiac output to carry oxygenated blood.
      iv. Provides better coronary blood flow.
      v. Can be done via cutdown or percutaneous.
   b. Veno-Arterial (VA).
      i. Placing separate cannulas into an artery and a vein (carotid, jugular, femoral, axillary).
      ii. Blood is withdrawn from a large vein and then returned to the arterial systemic circulation.
      iii. Can support the patient strictly through increasing flow through the circuit.
      iv. Provides better systemic circulatory support.
      v. Increases cardiac afterload due to the fluid and pressure returned to the systemic circulation.
6. Cannulation:
   a. Percutaneous:
      i. Performed at beside
      ii. Fluoroscopic assistance
      iii. Echocardiogram assistance
      iv. Internal jugular–carotid, femoral–femoral, jugular–femoral, jugular–axillary, axillary–femoral
   b. Open cutdown:
      i. Incision over desired vessel and expose
      ii. Similar options—jugular, carotid, femoral, axillary
   c. Hybrid:
      i. This is a way of using a modified cutdown along with percutaneous method to allow visualization while percutaneously gaining vascular access.
   d. Distal perfusion catheter:
      i. If concerned about distal extremity perfusion, can place a distal catheter in radial or posterior tibial vessel and retrograde flow.
      ii. Some institutions will place cephalad catheters as well with VV cannula to improve cerebral blood flow and oxygenation.

7. Anticoagulation—once the patient is cannulated and being run on the ECMO circuit, they will require anticoagulation to prevent clotting within the cannulas, tubing, or the circuit. Heparin is usually the agent of choice, but more centers are beginning to use direct thrombin inhibitors. Some circuits are being developed that have thromboresistant surfaces or agents coating the tubing to prevent clotting.
    a. Heparin—this agent is a non-specific anticoagulant and can be unpredictable.
        i. Centers will use activated clotting time (ACT) levels to determine their heparin level and adjust dosing on ACT ranges.
        ii. Another option is following anti Xa levels. This more specifically measures the indirect amount of Heparin in the bloodstream by measuring its inhibition of factor Xa activity. This has been shown to be more accurate and allows less heparin to be used, more stable levels and decreasing the amount of blood products used.
    b. Direct thrombin inhibitors (Argatroban, Bivalirudin, Lepirudin):
        i. Bind directly to thrombin to prevent the activation of clotting factors.
        ii. Can provide a more predictable anticoagulant effect.
        iii. Can cause less platelet activation.
        iv. Some centers are switching to full direct thrombin inhibitors as their agent of choice for the ECMO circuit.
8. Types of circuit:
    a. Roller pump:
        i. Used mainly in neonates.
        ii. Roller pump that pushes blood through the circuit.
        iii. Dependent on gravity drainage but less with the better bladder.
        iv. No feedback mechanism to stop the pump if distal obstruction in circuit, risk of rupture.
        v. Can flow at lower rates and cause less hemolysis.
    b. Centrifugal pump:
        i. Can be used in neonatal, pediatric, and adult ECMO (some programs using all centrifugal pumps).
        ii. Magnetic cone that circulates blood through the system, creates a negative pressure in the system, and “pulls” blood out of the patient.
        iii. Has the ability to sense high negative pressure or high circuit pressure and will slow down and even cut out the pump to prevent rupture.
        iv. Can flow from low to high rates depending on need. Some concern that higher risk of hemolysis at low flow due to more turbulent blood flow.

9. Physiologic goals:
    a. Improve tissue perfusion:
        i. Goal $SvO_2$ of 60–80%—measures the amount of oxygen left over in the circulation after delivering oxygen to the tissues.
        ii. Oxygen delivery:
            1. Arterial $O_2$ content ($CaO_2$) = $(1.36 \times Hb \times SaO_2) + (0.0036 \times PaO_2)$.
        iii. Oxygen consumption:
            1. Amount of $O_2$ used in metabolic processes.
            2. Independent of oxygen delivery until patient hypoxic.
            3. Muscular activity, hyperthermia, and increased catecholamines increase consumption.
            4. Rest, paralysis, and hypothermia slow down consumption.
        iv. Goals:
            1. $DO_2$:$VO_2$ ratio of 5:1.
            2. $DO_2 = CaO_2 \times (HR \times SV)$.
            3. Anaerobic metabolism occurs at $DO_2$:$VO_2$ ratio of 2:1.
10. Complications:
    a. Circuit:
        i. Clots:
            1. Clots can occur anywhere in the circuit.
            2. They usually form in low flow areas or areas where turbulence increases (change in size of caliber of tubing or circuit).
            3. Clots will form in the oxygenator and lead to failure.
            4. May require complete circuit change.
        ii. Rupture:
            1. Rare occurrence.
            2. Usually due to some high pressure in the circuit from clot or improper clamping of the circuit, loose connectors.
        iii. High pressures:
            1. Can occur in both venous and arterial limb.
            2. Usually related to clots in the circuit.
            3. Can be from patient hypovolemia and low blood volume.
        iv. Oxygenator failure:
            1. Can be due to manufacturer error.
            2. Usually due to high clot burden in circuit and oxygenator.

        v. Oxygenator leakage:
            1. Plasma leakage can be problematic in some of the oxygenators causing failure.
        vi. Air in circuit:
            1. Due to accessing the circuit and not clamping or inadequate tightening of components.
            2. Bubble detectors in place to detect.
            3. Will need to stop circuit and try to float air to remove or Centrifugal pump will capture air.
            4. Can be lethal if large amount of air gets into circuit and makes it to the patient.
    b. Patient:
        i. Bleeding:
            1. Can occur at any site—IV sticks, cannula sites, any surgical sites, spontaneous into chest, abdomen, or other cavities.
            2. Limit procedures and IV sticks.
            3. Risk of intracerebral hemorrhage causing IVH, stroke, or seizures.
            4. Highest risk with surgical procedures performed while patient on ECMO and anticoagulated.
        ii. Neurologic:
            1. Risk of intraventricular hemorrhage, stroke, or seizures.
            2. Long-term follow-up recommended, especially for neonates as high risk of neurologic impairment.
        iii. Renal:
            1. Renal injury common, usually reversible.
            2. Electrolyte abnormalities common, correctable with replacements.
            3. May require continuous veno-venous hemodialysis (CVVHD) to remove fluid and protect kidneys.
            4. Rarely will require long-term dialysis after coming off ECMO.
        iv. Loss of limb:
            1. If child is put on ECMO for sepsis, risk of micro emboli and clots lodging in peripheral vessels leading to digit and even limb ischemia.
            2. Rarely requires amputation.
            3. Circuit clots can dislodge and float to head or extremities causing ischemia.

11. Outcomes: international outcomes are measured and tracked by the Extracorporeal Life Support Organization (ELSO). This organization was started in the late 1980s out of Ann Arbor where they developed a standardized data sheet to follow outcomes as well as complications. Reports are sent out

twice a year and centers can follow their outcomes and compare them to international data. The number of international runs since 1990 are 133,371, out of these runs 93,776 survived ECLS (70%) and 73,913 (55%) survived to discharge from the hospital. These numbers include neonatal, pediatric, and adult as well as pulmonary, cardiac, and ECPR cases. For neonatal pulmonary cases, survival with ECLS is 87% and survived to discharge from hospital is 74%. These numbers decrease for pediatric and adult pulmonary populations. For pediatric population, survival with ECLS is 72% and survival to discharge is 60%. For adults, survival with ECLS is 69% and survival to discharge is 60%.

## Additional Notes

ECMO has a history interwoven with cardiac bypass. Dr. Gibbon and his wife developed an oxygenator after the death of a patient from a pulmonary embolus. They felt there was a better way to manage patients to prevent death. The first use of an oxygenator was in 1953 on a patient with an ASD that was repaired. From this start, clinical trials began in the 1960s. The first survivor with the use of an oxygenator was in 1971 by Dr. J Donald Hill in a patient who suffered a ruptured aorta from a motorcycle crash. In 1972, Dr. Robert Bartlett at the University of California Irvine was the first to use a membrane oxygenator in a neonate with meconium aspiration that survived. He then published articles on the survival of ECMO in neonates and its use increased. Over the past 40 years, the need for ECMO in neonates has decreased due to advances in critical care. However, its use has increased in the pediatric and adult populations, especially in cardiac uses. Other uses for ECMO have been discovered as well as new technologies. ECMO can be used for $CO_2$ removal alone, ECPR, used for warming hypothermic patients, trauma patients with ARDS, bridge to transplant and donation, ambulatory ECMO, and development of artificial placentas.

## Study Questions

1. Which of the following is NOT a contraindication to ECMO in a newborn?
    a. Intracerebral hemorrhage
    b. Pulmonary hemorrhage
    c. Weight <2000 grams
    d. Prematurity <32 weeks gestation
    e. Chromosomal abnormality

Answer:

(b.) Pulmonary hemorrhage is not a contraindication to ECMO in a newborn. Intracerebral hemorrhage is a contraindication because of the need for anticoagulation. Prematurity is a contraindication due to increased risk for intraventricular hemorrhage. Weight less than 2000 g is a contraindication because of the size of the vessels relative to the minimum necessary size of the cannulas although ECMO has been used in patients weighing as little as 1800 g. Irreversible congenital conditions including chromosomal abnormalities are a contraindication. Pulmonary hemorrhage is not a contraindication, but care must be taken with anticoagulation.

2. A full-term baby with congenital diagnosis that requires ECMO is discharged on gastrostomy feeds. All the following are increased in incidence compared to critically ill newborns not requiring ECMO EXCEPT:

   a. Impaired hearing
   b. Risk of autism spectrum disorder
   c. Attention deficit hyperactivity disorder (ADHD)
   d. Delayed growth
   e. Decrease in overall intelligence

   Answer:

   (e.) Decrease in overall intelligence is not increased in incidence. Newborns that undergo ECMO are critically ill. ADHD and autism spectrum disorder are approximately ten times more common in children that underwent ECMO for CDH. Sensorineural hearing loss can be as high as 50% but may be exacerbated by aminoglycosides and furosemide diuretic. There is delayed growth in the first 2 years of life. Overall intelligence has not been demonstrated as decreased compared to matched controls.

3. Which of the following criteria is not an indication to place a child in pulmonary failure on ECMO?

   a. Oxygenation index >40
   b. Alveolar-arterial gradient > 610 on two consecutive blood gases
   c. Respiratory alkalosis
   d. $PaO_2/FiO_2$ ratio <100 mm Hg

   Answer:

   (c.) Respiratory alkalosis is not an indication to place a child on ECMO. Children with severe pulmonary failure will have respiratory acidosis, not alkalosis. ECMO is reserved for children with severe but potentially reversible pulmonary failure when all medical therapy has been exhausted. There are several criteria that can be measured and calculated to help decide when the pulmonary failure is severe enough to warrant ECMO. The oxygenation index (OI) is calculated by the (Mean Airway Pressure (MAP) × $FiO_2$ × 100) / $PaO_2$. An OI > 40 is an indication for ECMO. For an OI > 40, mortality is 80%, and for an OI of 25–40 the mortality is 50%. The alveolar–arterial gradient (A-a Gradient) is calculated as ($FiO_2$ (713) − $PaO_2$ − $PaCO_2$)/$PaO_2$. An A-a Gradient <610 on at least two consecutive blood gases is an indication for ECMO and carries a 79% mortality. The P/F ratio is calculated as $PaO_2/FiO_2$ and a P/F ratio < 100 mm Hg is an indication for ECMO. All these criteria are an attempt to quantify the degree to which the pulmonary failure is impeding elimination of carbon dioxide and serving as a barrier for oxygenation to cross from the airway to the bloodstream.

4. What is an advantage of Veno-Venous (VV) ECMO over Veno-Arterial (VA) ECMO?
    a. Provides better systemic circulatory support.
    b. Provides better coronary blood flow.
    c. Is not dependent on cardiac output to carry oxygenated blood.
    d. Increases cardiac afterload due to the fluid and pressure returned to the systemic circulation.

Answer:

(b.) Provides better coronary blood flow. VV ECMO is delivered through a venous cannula (and sometimes multiple venous cannulas) so that blood is both withdrawn and returned to the venous compartment of the bloodstream. VV ECMO returns oxygenated blood to the vena cava and the right heart. It is dependent on natural cardiac output to carry this oxygenated blood to the rest of the body. VV ECMO provides better coronary blood flow. VA ECMO involves placing separate cannulas into an artery and a vein. VA ECMO can directly support systemic circulation through increasing flow through the circuit. VA ECMO also increases cardiac afterload because the output of blood is returned to the systemic circulation downstream from the aortic valve.

## Further Reading

Annich G, Lynch W, MacLaren G, Wilson J, Bartlett R. Extracorporeal cardiopulmonary support in critical care. fourth ed. Ann Arbor, MI: Extracorporeal Life Support Organization; 2012.

Bailly DK, Reeder RW, Zabrocki LA, Hubbard AM, Wilkes J, Bratton SL, Thiagarajan RR. Development and validation of a score to predict mortality in children undergoing ECMO for respiratory failure: pediatric pulmonary rescue with extracorporeal membrane oxygenation prediction (P-PREP) score. Cri Care Med. 2017;45(1):e58–66.

Barbaro RP, Boonstra PS, Paden ML, Roberts LA, Annich GM, Bartlett RH, Moler FW, Davis MM. Development and validation of the pediatric risk estimate score for children using extracorporeal respiratory support (Ped-RESCUERS). Intensive Care Med. 2016;42(5):879–88.

Extracorporeal life support registry report. https://www.elso.org/Registry/Statistics/InternationalSummary.aspx. Accessed July 2020.

Kim ES, Stolar CJ. ECMO in the newborn. Am J Perinatol. 2000;17(7):345–56.

Makdisi G, Wang IW. Extra orporeal membrane oxygenation (ECMO) review of a lifesaving technology. J Thoracic Disease. 2015;7(7):E166–76.

Nellis ME, Vasovic LV, Goel R, Karam O. Epidemiology of the use of hemostatic agents in children supported by extracorporeal membrane oxygenation: A pediatric health information system database study. Front Pediatr. 2021;9:673613.

Padhya DR, Prutsky GJ, Nemergut ME, Schears GS, Flick RP, Farah W, Wang Z, Prokop LJ, Murad MH, Alswas M. Routine laboratory measures of heparin anticoagulation for children on extracorporeal membrane oxygenation: systematic review and meta-analysis. Thromb Res. 2019; Online Publication.

Skinner SC, Hirschl RB, Bartlett RH. Extracorporeal life support. Semin Pediatr Surg. 2006;15(4):242–50.

Stocker CF, Horton SB. Anticoagulation strategies and difficulties in neonatal and paediatric extracorporeal membrane oxygenation (ECMO). Perfusion. 2016;31(2):95–102.

Valencia E, Nasr G. Updates in pediatric extracorporeal membrane oxygenation. J Cardiothorac Vasc Anesth. 2020;34(5):1309–23.

Van Ommen CH, Neunert CE, Chitlur MB. Neonatal ECMO. Front Med. 2018;5:289.

# Pyloric Stenosis

**Marc S. Lessin**

**Key Points**

1. Pyloric stenosis is the most common surgical cause of vomiting in infancy.
2. The vomiting is always non-bilious and is forceful or "projectile," typically after oral intake. Vomiting occasionally may be rust colored from blood.
3. The gold standard for diagnosis is ultrasound of the pyloric muscle dimensions.
4. Dehydration should be suspected and electrolytes corrected with intravenous fluids.
5. Treatment is pyloromyotomy (open or laparoscopic).

## 1. Background

Pyloric stenosis is a congenital condition which is caused by hypertrophy of the pyloric sphincter muscle. The hypertrophy causes a relative obstruction of gastric emptying which results in forceful or projectile post-prandial vomiting. The diagnosis is suspected by history and typically confirmed by ultrasound of the stomach focusing on the pyloric area. Treatment is a pyloric muscle splitting operation known as pyloromyotomy after correction of any electrolyte abnormalities, particularly bicarbonate.

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2. Pathophysiology

   a. Epidemiology: The incidence of pyloric stenosis has been increasing. It is estimated to occur in 1 out of every 250 live births. It is more common in first-born males with a male-to-female incidence of 4:1. The typical presentation is between 2 and 8 weeks of age. Pyloric stenosis rarely occurs after 4 months of age but may present later in premature infants. There is a familial predisposition with approximately 1 in 20 male offspring and 1 in 50 female offspring from an affected male parent. The incidence of pyloric stenosis in a first-born male from an affected female parent has been reported as high as one in five.
   b. Etiology: The etiology of pyloric hypertrophy is unknown. No genetic factors have been identified but there is a familial predisposition. Risk factors include family history, young maternal age, and erythromycin exposure prenatally or in the newborn period.

      In pyloric stenosis, the pyloric muscle is thickened. The muscular hypertrophy develops after several weeks of life. The hypertrophy results in narrowing of the pyloric lumen which eventually causes near-total obstruction of the gastric outlet. The stomach increases in size. The thickened pyloric muscle protrudes distally into the duodenal lumen which can be noted as the "shoulder sign" on upper gastrointestinal contrast study, which is rarely performed as sonography is diagnostic. The redundant mucosa of the duodenum with the overlapping pyloric muscle is a "danger zone" during pyloromyotomy as extension of the pyloromyotomy can result in violation of the duodenal mucosa.

3. Clinical presentation

   Infants with pyloric stenosis typically present at 2–8 weeks of age. Premature infants account for about 10% of cases and typically present at 42–50 weeks post-conceptual age. Presentation beyond 3 months is unusual accounting for less than 5% of cases and presentation beyond 4 months does not occur.

4. Diagnosis

   a. Symptoms: The typical symptom is post-prandial, forceful, or "projectile" non-bilious vomiting in an infant in the expected age group. The vomiting typically occurs 30–60 min after taking a bottle. The infant is typically hungry after eating and eager to eat as opposed to gastroesophageal reflux symptoms in which infants may be disinterested after vomiting due to associated discomfort with emesis. Often, there have been one or more formula changes in an attempt to alleviate the vomiting. If the vomiting has gone on for more than several days, there may be decreased urine, weight loss, and dehydration.
   b. Examination: The abdomen may be distended due to gastric dilation and become scaphoid after an episode of emesis. Gastric peristaltic waves may be seen across the abdomen after a feed. The hypertrophic pylorus can be palpated in the right upper quadrant. It is best palpated by placing a gastric

decompression tube and giving the baby a bottle of a dextrose water solution. The hypertrophic pylorus can be palpated in the right upper quadrant beneath the liver edge. It is best palpated by sweeping down from beneath the liver edge. The pylorus feels like an elongated mobile firm structure described as an "olive pit." The caudate lobe of the liver, right kidney, vertebrae, and gastric decompression tube can be mistaken for the hypertrophied pylorus. Physical examination is rarely if ever the sole indication for surgical intervention. In practice, the diagnosis of pyloric stenosis is almost exclusively made on ultrasound often before evaluation by the surgeon.

c. Laboratory tests: The only required laboratory evaluation is electrolytes. Most will also obtain a complete blood count and fractionated bilirubin if jaundice is present. Most infants will have normal electrolytes at the time of presentation. The common electrolyte abnormality is hyponatremic, hypochloremic, and hypokalemic metabolic alkalosis. Occasionally children will present with paradoxical aciduria. The electrolyte abnormalities are explained by the ensuing dehydration due to vomiting which results in resorption of sodium in exchange for potassium at the renal tubule. As potassium becomes depleted, the aldosterone mechanism results in resorption of potassium at the expense of the hydrogen ion being excreted in the urine. Elevated bilirubin present in 10% is due to elevated indirect component which is due to decreased activity of glucuronyl transferase which conjugates bilirubin. This will resolve once the infant is able to feed.

d. Radiographic studies: Ultrasound is the gold standard for diagnosis of pyloric stenosis with a sensitivity of 97%. In pyloric stenosis, the wall thickness (radius) is ≥3 mm and the pyloric channel length is >14 mm (Fig. 1). The radiologist will often have the infant take a dextrose solution to determine if anything passes through the pyloric channel and to exclude pylorospasm which can give a false-positive study. Occasionally, the diagnosis is made on

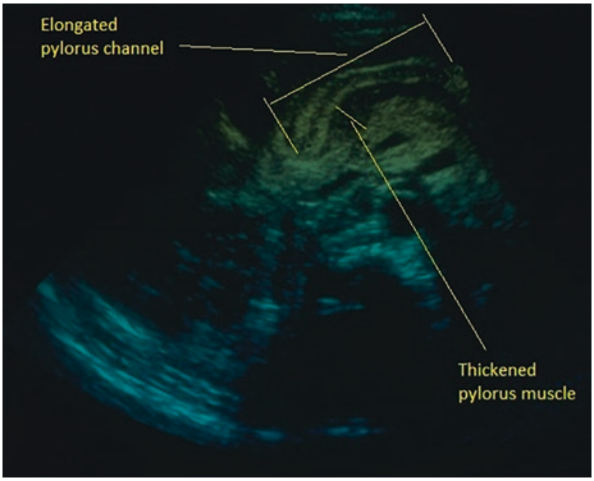

**Fig. 1** Typical appearance on ultrasound of pyloric stenosis. Note the mucosal thickening, elongated pyloric channel, and wall thickness (radius) 3 mm. (Source: Marc S. Lessin, MD)

fluoroscopic upper gastrointestinal contrast study which will show a wisp of contrast passing through the narrowed pyloric, the so-called "string sign," and/or impression of the pylorus indenting the stomach, the so-called "shoulder sign."

5. Treatment
   a. Initial measures: Electrolytes must be checked for the common disturbances of hyponatremia, hypochloremia, hypokalemia, and alkalosis. A gastric decompression tube should not be placed as this increases fluid losses and loss of gastric acid. A fluid bolus should be initiated with 10–20 ml/kg of normal saline.
   b. Subsequent measures: The infant should be made NPO. Intravenous fluids should be initiated at 25%–50% above maintenance. Mild to moderate electrolyte abnormalities can be corrected with intravenous fluids of 0.45% normal saline with 5% dextrose with 20 mEq/L potassium. Potassium should be withheld until the baby has urinated. In cases of severe dehydration, 0.9% normal saline with 5% dextrose may be necessary to correct dehydration. When administering normal saline, electrolytes should be checked frequently. Potassium should be added to the intravenous fluids even if values are normal as most potassium is intracellular.
   c. Preoperative preparation: The surgical procedure is not emergent. Electrolytes should be corrected before surgical intervention. In particular, serum bicarbonate should be less than 30 mEq/L because of respiratory depression which will impair ability to extubate after the surgical procedure and can cause postoperative apnea. In severe cases, correction of electrolytes may take several days. Preoperative antibiotics are unnecessary as the case is a "clean case."
   d. Operative treatment: The treatment is extramucosal pyloromyotomy. The procedure is done under general endotracheal anesthesia. The operation can be performed open or laparoscopically. The vast majority of pyloromyotomies performed by pediatric surgeons are performed laparoscopically. No distinct advantage has been shown for the laparoscopic approach other than cosmetic concerns. The laparoscopic approach involves three incisions: one trocar placement at the umbilicus for the camera and two in the upper abdomen through which 3 mm instruments are directly inserted. The right upper quadrant incision site is used to stabilize the pylorus and a scalpel or electrocautery inserted in the left upper quadrant to open the serosa. A laparoscopic spreader is then exchanged through the left upper quadrant site to split the circular and longitudinal muscles. The anesthesiologist then insufflates air into the stomach to test for a leak.

      The open approach is most commonly performed through either a right upper quadrant transverse incision or a supraumbilical curvilinear incision. The stomach is located and the hypertrophic pylorus gently delivered through the incision. The serosa is scored with electrocautery, and a specialized pyloromyotomy spreader (or handle of the scalpel blade) is used to split the muscle fibers. The pyloromyotomy is tested by inserting a finger into the

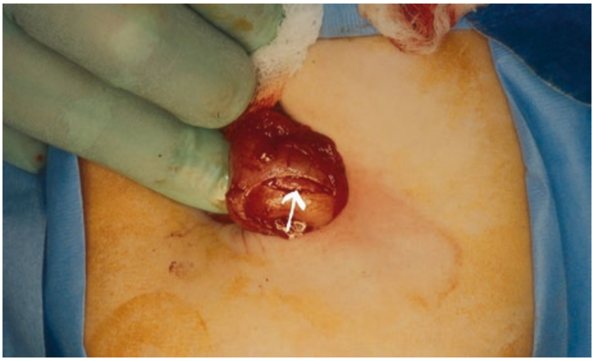

**Fig. 2** Operative pyloromyotomy through a supraumbilical incision. Arrow points to the bulging mucosa. (Source: Marc S. Lessin, MD)

duodenum and recognizing a break in the pyloric ring and insuring that both sides rock independently. The stomach can gently be compressed to assess for a leak. The pylorus is then returned to the peritoneal space and the incision closed in layers (Fig. 2).

In both procedures, the incision is made between the vascular branches to the greater and lesser curvature of the stomach to avoid bleeding. The incision is carried within 1–2 mm of the pylorus identified by a color change and the vein of Mayo which should not be crossed. The proximal extension of the pyloromyotomy should extend onto the antrum of the stomach where the hypertrophied muscle dissipates.

e. Postoperative treatment: Infants should be monitored for apnea following the procedure done under general anesthesia. Acetaminophen is usually all that is needed for pain control. Most post-pyloromyotomy feeding regimens involve a short period of nothing by mouth until the baby awakens and initial low-volume formula feeds every 3 h for 1–2 feeds then ad libitum feeds. Some surgeons prefer a regimented feeding schedule. Vomiting after pyloromyotomy is common and usually resolves in the first 48 h. The usual postoperative length of stay is 24–36 h.
f. Complications: Complications after pyloromyotomy are rare. Persistent vomiting beyond 48 h may be the result of incomplete pyloromyotomy. Incomplete pyloromyotomy may be slightly more common with the laparoscopic approach. This is best evaluated by upper gastrointestinal fluoroscopic studies as ultrasound post-pyloromyotomy is difficult to interpret as hypertrophy of the muscle may still be present. Occasional bleeding from the muscular edge may occur during the pyloromyotomy which can be controlled with electrocautery. Incisional hernias or hernias at laparoscopic incisions have been reported. Wound infections are uncommon. Perforations of the mucosa can be repaired with a fine absorbable suture if on the margins of the pyloromyotomy and bolstered with omentum. Central perforations require closure of the pyloromyotomy, rotation of the pylorus 180° and performing a pyloromyotomy on the posterior surface of the pylorus. If a perforation requiring a new myotomy occurs during the laparoscopic approach, the

procedure should be converted to open. For perforations, a nasogastric tube should be placed after repair of a perforation with delay in feeding of 24 h. A water-soluble contrast study is usually performed to confirm the integrity of the repair before feeds.

## Additional Notes

It was originally believed the condition was the result of pylorospasm. In the early part of the twentieth century, treatment including antispasmodics and various feeding regimens such as frequent small volume, low curd feeds were used. Mortality was high but those that survived would eventually outgrow the condition. It was speculated that those that survived may have suffered from pylorospasm rather than pyloric stenosis. The condition was first described in the 1700s and later by the famous pathologist Harald Hirschsprung in 1888. Extramucosal pyloromyotomy was revolutionary insofar as survival from this condition was described by Conrad Ramstedt in 1912. The operation still follows the basic tenets described in the original paper.

## Study Questions

1) Which of the following is the most common electrolyte disorder in pyloric stenosis:

   A. Hyponatremic, hypochloremic, hypokalemic metabolic alkalosis.
   B. Hyponatremic, hypochloremic, hyperkalemic metabolic alkalosis.
   C. Hypernatremic, hyperchloremic, hypokalemic metabolic alkalosis.
   D. Hypernatremic, hyperchloremic, hypokalemic metabolic acidosis.
   E. Hyponatremic, hypochloremic, hyperkalemic metabolic acidosis.

Answer: A: The persistent vomiting in pyloric stenosis initially results in loss of fluids and dehydration with loss of sodium chloride. The body's initial response to dehydration is to preserve sodium. This results in loss of potassium chloride at the renal tubule. The loss of gastric contents with its low pH results in alkalosis. The increased sodium bicarbonate results in the metabolic alkalosis and is important to correct prior to surgery as it causes respiratory depression and postoperative apnea.

2) A 4week-old presents with a 1-week history of forceful vomiting. The baby weighs 4 kg. Ultrasound shows pyloric stenosis. Electrolytes show sodium = 132 mEq/L, potassium = 3.2 mEq/L, chloride = 89 mEq/L, carbon dioxide = 35 mEq/L. A 20 ml/kg fluid bolus of normal saline is given. Which is the most appropriate maintenance fluid and rate?

   A. D5 0.225% normal saline solution with 10 mEq/L K+ at 20 ml/h.
   B. D5 0.9% normal saline solution @ 24 ml/h.

C. D5 0.45% normal saline solution with 20 mEq/L KCl at 20 ml/h.
D. D5 0.9% normal saline solution with 40 mEq/L KCl @ 24 ml/h.

Answer: C: The electrolytes demonstrate moderate dehydration. D5 0.225 normal saline is never appropriate for preoperative resuscitation in a baby with pyloric stenosis. Normal saline (0.9%) may be appropriate in cases of severe dehydration but must be administered cautiously with frequent electrolytes checks to avoid fluid overload. In the scenario given, the most appropriate solution D5 0.45% normal saline with 20 mEq KCl/L.

3) Studies have shown a clear advantage of laparoscopic over open pyloromyotomy for which of the following?

A. Lower incidence of incomplete pyloromyotomies.
B. Shorter hospital stay.
C. Less major complications.
D. Better cosmesis.

Answer: D: Studies have shown that outcomes of laparoscopic versus open pyloromyotomy are similar. There may be a slight increase in incomplete pyloromyotomies with the laparoscopic approach. Time to full feeds is slightly shorter but did not result in shorter hospital stay. Reported complications are about the same for both approaches. There is clearly improved cosmesis with the laparoscopic approach compared to an open technique incision.

## Further Reading

Adibe OO, Igbal CW, Sharp SW, Juang D, et al. Protocol versus ad libitum feeds after laparoscopic pyloromyotomy: a prospective randomized trial. J Pediatr Surg. 2014;49(1):129–32.

Ashby HT, Southam AH. The early diagnosis and treatment of pyloric stenosis in infants. Br Med J. 1925;1:403–5.

Costanzo CM, Vinocur C, Berman L. Postoperative outcomes of open versus laparoscopic pyloromyotomy for hypertrophic pyloric stenosis. J Surg Res. 2018;224:240–4.

Sathya C, Wayne C, Gotsch A, Vincent J, et al. Laparoscopic versus open pyloromyotomy in infants: a systematic review and meta-analysis. Pediatr Surg Int. 2017;33(3):325–33.

Taghavi K, Powell E, Bhaveshkumar P, McBride CA. The treatment of pyloric stenosis: evolution in practice. J Paediatr Child Health. 2017;53(11):1105–10.

# Duodenal Atresia

**Samir Narula, Pratibha Vemulapalli, and Brian Gilchrist**

**Key Points**

1) Newborns with intolerance of feeds and vomiting may have a congenital obstruction of the duodenum due to atresia or stenosis, a condition strongly associated with Trisomy 21.
2) The hallmark of diagnosis is the double-bubble pattern of gas seen on abdominal radiographs reflecting the stomach and a proximally dilated duodenum.
3) Prognosis after treatment with a duodenoduodenostomy is excellent, with survival rates exceeding 95%.

1) Epidemiology
    a) Incidence:

Intrinsic duodenal obstruction (DO) caused by atresia or stenosis is the most common cause of congenital intestinal obstruction and is estimated to occur in 1 per 5000 to 10,000 live births, affecting males more commonly than females with a 2:1 ratio. Approximately half of infants with DO have associated congenital anomalies, particularly Trisomy 21 (Down syndrome) which is present in up to 50% of cases. While yet to be discovered, the strong association between DO

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and Down syndrome as well as several reports of familial cases of DO suggest an underlying genetic etiology. Other anomalies found in high frequency with DO include annular pancreas, congenital heart disease, malrotation, preduodenal portal vein, esophageal atresia, anorectal anomalies, and renal/urinary tract anomalies.

b) Etiology:

Congenital duodenal obstruction is the result of embryologic defects in foregut development, recanalization, or rotation. The resultant obstructing lesions are classified as either intrinsic or extrinsic. Intrinsic lesions include duodenal atresia, duodenal stenosis, or duodenal web/diaphragm. Lesions classified as extrinsic include annular pancreas, malrotation, peritoneal bands, and anterior portal vein.

Whereas intestinal atresia distal to the duodenum is the consequence of intrauterine vascular accidents, atresia of the duodenum is unique and due to failure of recanalization. Although the underlying cause of DO is unknown, its embryologic pathophysiology was first theorized by Julius Tandler in 1900 and has been well described since. *In utero* the endoderm gives rise to the gut tube during the fourth week of development. During the sixth week, the gut epithelium rapidly proliferates causing complete occlusion of the intestinal lumen. Over the course of the eighth through tenth week, epithelial apoptosis occurs, gradually recanalizing and re-establishing continuity of the now epithelium-lined intestinal lumen. Failure to recanalize or incomplete recanalization leads to DO.

2) Pathophysiology

a) Anatomy:

The spectrum of malformations responsible for DO ranges from webs (with or without fenestration) of variable thickness in a duodenum that is in continuity to complete duodenal discontinuity and can be classified as causing complete or incomplete obstructions. The lesions can also be classified as pre-ampullary or post-ampullary based on their location relative to the ampulla of Vater, with nearly 85% of lesions being post-ampullary. Incomplete obstructions are considered stenoses and are due to a fenestrated web or diaphragm. As stenosis may allow for the flow of some succus, it may not become apparent until later when solid foods are introduced. Stenosis most commonly involve the third and/or fourth segment of the duodenum. Complete obstructions or atresias generally occur near the ampulla of Vater and are further classified into three distinct morphologic subtypes as originally described by Gray and Skandalakis (Fig. 1).

I. Type I atresias contain an obstructing luminal membrane (web/diaphragm) of varying thickness composed of mucosal and submucosal layers. These are the most common, accounting for >90% of all intrinsic duodenal obstructions. Occasionally the web may elongate or balloon distally known as a "windsock deformity". This variation is important to understand as the origin of the web is more proximal than the dilated duodenum, usually the

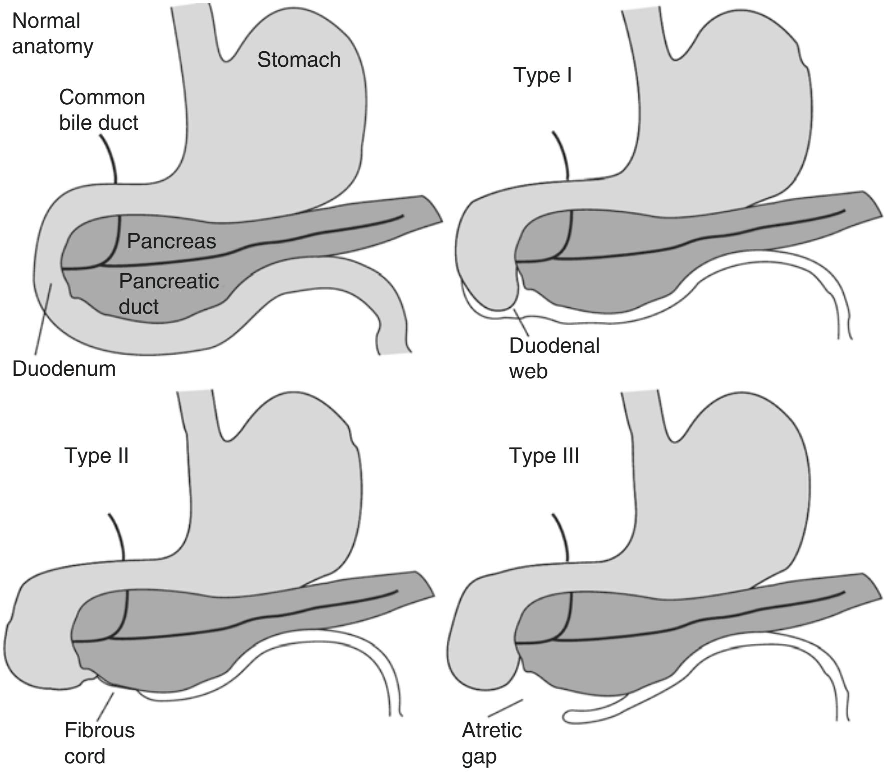

**Fig. 1** Classification of duodenal atresia. (Source: Coppola C.P. (2014) Duodenal Atresia. In: Coppola C., Kennedy, Jr. A., Scorpio R. (eds) *Pediatric Surgery: Diagnosis and Treatment*. Springer, Cham. https://doi.org/10.1007/978-3-319-04340-1_23).

second portion of the duodenum. The membrane may have a central fenestration allowing some succus passage, with the degree of obstruction and symptoms being inversely proportional to the diameter of the opening.

II. Type II atresias contain two atretic ends of duodenum connected by a short fibrous cord without a defect in the mesentery.
III. Type III atresias are completely discontinuous with a gap between the atretic proximal and distal duodenal segments with an associated mesenteric defect.

3) Prenatal diagnosis

Duodenal atresia is diagnosed prenatally in up to 60% of cases. Two major sonographic findings are suggestive of duodenal obstruction: polyhydramnios and the “double-bubble” sign. Maternal polyhydramnios is reported in 30–60% of cases. The classic “double-bubble” sign represents an obstructive pattern of simultaneous gastric and proximal duodenal dilation, with the larger of the two representing the dilated fluid-filled stomach and the other the dilated proximal duodenum. The two “bubbles” are separated by narrowing associated with pylorus. Most prenatal sonographic diagnoses occur in the third trimester likely owed to the hydrostatic pressure necessary to dilate the duodenum; however, rare reports of diagnosis as early as 12 gestational weeks exist.

4) Clinical presentation and diagnosis

   a) Symptoms:

The clinical presentation is consistent with a proximal intestinal obstruction, with vomiting being the most common symptom and often presents within the first 24 h of life. Given the spectrum of disease, the clinical features manifest according to the degree of and site of obstruction. Most lesions (85%) are post-ampullary and result in bilious emesis, while those proximal to the ampulla result in non-bilious emesis. In some cases, meconium is passed followed by constipation and does not exclude the diagnosis. In those with an incomplete obstruction secondary to duodenal stenosis, the presentation may be delayed until childhood or in extremely rare cases adulthood.

b) Examination:

Abdominal distention is generally minimal or not observed because of the proximal location of the obstruction. Epigastric fullness representing the dilated stomach and proximal duodenum may be appreciated. Passage of a gastric tube yields air and fluid that may be bilious or non-bilious.

c) Laboratory tests:

Lab work may reveal hypoglycemia and electrolyte imbalances due to loss of gastric, pancreatic, and biliary secretions via emesis. Karyotype analysis should be performed due to the strong association with Trisomy 21.

d) Radiographic studies:

Imaging including prenatal sonogram and plain abdominal radiograph is essential to diagnosis. Equivocal cases may require upper gastrointestinal contrast series (UGIS) for diagnosis. In those without prior nasogastric (NG) or orogastric (OG) tube decompression, erect radiographs will show the typical “double-bubble” sign as seen in Fig. 2. The characteristic “double-bubble” reflects gastric dilation and a dilated proximal duodenum. Those who have previously undergone NG tube decompression may require an injection of air via NG tube to yield a clearer “double-bubble” sign. In cases of duodenal stenosis and webs with fenestration, X-ray will show scant air distal to the duodenum. Air may be seen in rare cases distal to the obstruction in those with duodenal atresia due to a bifid common bile duct (CBD) with one limb proximal and one limb distal to the obstruction.

A UGIS can be a useful adjunct to aid in diagnosis in unclear presentations or to confirm duodenal stenosis or fenestrated duodenal webs in cases where scant air is seen distal to the site of obstruction. It is also used to differentiate other causes of duodenal obstruction such as malrotation, volvulus, or duodenal duplications.

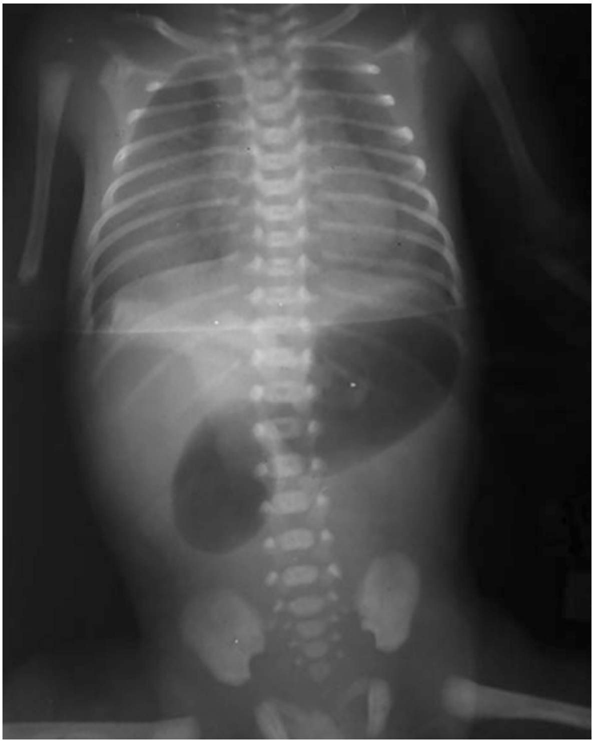

**Fig. 2** The larger of the two structures is the dilated stomach and the smaller bubble represents the dilated duodenum. (Source: Marc Lessin).

Echocardiogram should be performed prior to treatment to determine if an anatomic heart defect is present. Duodenal atresia has a strong association with Trisomy 21, and this carries a risk of congenital heart disease, particularly endocardial cushion or ventricular septal defects.

5. Treatment

   a) Initial measures:

   Initial management of DO requires admission to the neonatal intensive care unit (NICU) and basic laboratory tests including complete metabolic profile and complete blood count. Infants should undergo gastric decompression via OG or NG tube, fluid resuscitation, and electrolyte correction. Emergent surgery should only be performed in cases where concomitant volvulus cannot be excluded.

   b) Subsequent measures:

   Placement of a peripherally inserted central catheter (PICC) line to initiate parenteral nutrition should occur early in the course as enteric feeding is often delayed for several weeks from the time of diagnosis. Given the high frequency of other associated congenital anomalies in cases of DO, further evaluation using echocardiography and complete abdominal ultrasonography are essential to detect the presence of congenital heart defects, annular pancreas, and urinary tract or renal anomalies. In very premature infants, operative intervention should be delayed until pulmonary status and weight are more ideal.

c) Operative treatment:

In almost all situations, the best corrective option is to bypass the obstruction via duodenoduodenostomy. This is generally performed open through a transverse upper abdominal laparotomy. The entire bowel should be evaluated for malrotation, volvulus, as well as other atresias, stenoses, or anomalies. The duodenum is mobilized delicately, ensuring no injury to the ampulla of Vater or CBD. The NG or OG tube is then passed distally from the stomach until it stops signifying the location of obstruction or causing a dimpling of the duodenal wall as may be seen when a duodenal web is pulled distally by the tube in patients displaying a “windsock deformity”. When an annular pancreas is identified as the cause of obstruction or in addition to DO, it should never be resected or divided; bypass via duodenoduodenostomy is always the procedure of choice.

Before creating the duodenoduodenostomy, a Foley catheter should be passed proximally into the stomach and distally into the jejunum and pulled back with the balloon inflated to ensure distal luminal patency. Alternatively, the catheter may be passed directly into the open duodenum and saline infused to assess patency. The proximal dilated duodenum is then anastomosed to the decompressed distal duodenum most commonly using a “diamond-shaped” (Kimura) duodenoduodenostomy. This is created by making a transverse incision on the proximal duodenum and a longitudinal incision on the distal duodenum as shown in Fig. 3. The two incisions are positioned perpendicularly so that the anastomosis is held open to encourage the flow of gastric contents. Although rarely necessary, a tapering duodenoplasty may be performed when the proximal duodenum is grossly dilated to improve postoperative peristalsis. Some surgeons pass a transanastomotic nasojejunal tube at the time of operation to allow for early initiation of enteral feeding.

If performing a duodenoduodenostomy is challenging due to anatomic limitations such as atresia or stenosis located in the more distal third and fourth portions of the duodenum, the next choice is duodenojejunostomy, followed by gastrojejunostomy. However, these operations carry the risk of complications of blind loop with bacterial overgrowth and marginal ulceration; thus, they are considered inferior and should be avoided if possible.

In cases where a duodenal web is identified, an alternative approach to bypassing the obstruction is employed by some surgeons. The duodenum can be opened longitudinally and the web excised beginning laterally. Care should be taken to avoid injury to the ampulla of Vater when proceeding medially as it may form a portion of the web. In cases of involvement, it is wise to leave a rim of the medial wall surrounding the ampulla intact rather than risk injury. Once excised, the duodenum is closed transversely to prevent stricture formation. Given the risk of injury to the CBD and ampulla, many prefer to treat duodenal webs with duodenoduodenostomy. In recent years, endoscopic excision of duodenal webs has been performed with some success.

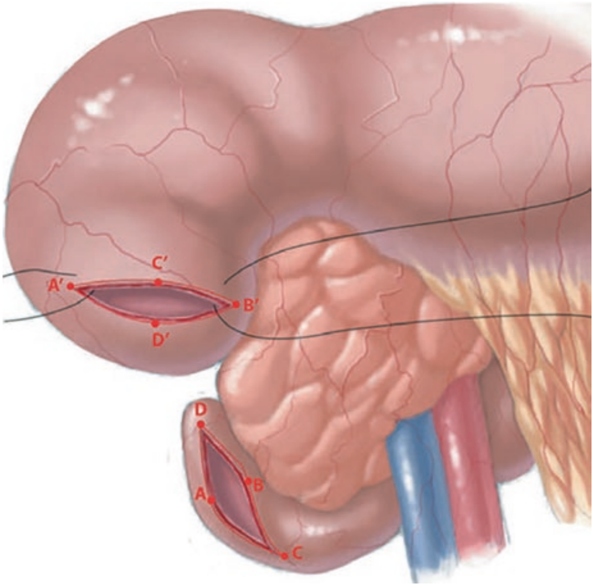

**Fig. 3** A transverse incision is made in the distal end of the proximal duodenum and a longitudinal incision is made in the duodenum distal to the occlusion. (Source: Sweed Y., Yulevich A. (2019) Duodenal Obstruction. In: Puri P. (eds) Pediatric Surgery. Springer, Berlin, Heidelberg. https://doi.org/10.1007/978-3-642-38482-0_63-2).

d) Postoperative treatment:

Postoperatively the baby should be returned to the NICU and continue to receive parenteral nutrition or a dextrose/saline infusion. Gastric decompression with an NG or OG tube should continue and fluid and electrolyte losses appropriately supplemented. Prolonged ileus is common due to ineffective peristalsis of the dilated proximal duodenum. The tube may be withdrawn, and oral feeding initiated once the gastric aspirate decreases in volume and bilious character, which may take up to three weeks.

e) Potential complications:

In the absence of specific concerns regarding the initial procedure, if relatively normal gastrointestinal function has not returned in three weeks, further evaluation is indicated. An UGIS should be obtained to evaluate for a residual or missed obstruction, anastomotic stenosis, or megaduodenum. Megaduodenum is a persistently dilated duodenum incapable of sufficient peristalsis; a tapering duodenoplasty is the procedure of choice to improve motility. However, any additional surgical procedure must be approached with caution as radiographic evidence of a dilated proximal duodenum may persist for several months in patients with an appropriately functioning duodenoduodenostomy.

Late complications occur in approximately 10% of patients and include delayed gastric emptying, severe gastroesophageal reflux, peptic ulcer disease, gastritis, blind loop syndrome, and obstruction secondary to adhesions as may occur following any intra-abdominal operation. Late duodenal obstructions may also occur secondary to fibrous food or a foreign body impacting at the location of a partially resected web, a strictured anastomosis, or an anastomosis that failed to appropriately grow with the child.

The early postoperative mortality rate is approximately 3–5% and is largely due to prematurity, low birth weight, or other associated congenital anomalies (mainly severe congenital heart disease). In comparison to 50 years ago when survival rates were roughly 45%, long-term outcomes are now excellent, with survival exceeding 95%. This can be attributed to a variety of factors, primarily improvements in NICU care, parenteral nutrition, management of associated congenital anomalies, and early prenatal diagnosis.

## Summary/Narrative

Since the first report of successful open repair of duodenal atresia by William Ladd in 1931, surgical techniques have advanced remarkably. Correction by laparotomy and diamond-shaped duodenoduodenostomy as described by Ken Kimura et al. in 1990 is the standard procedure with outstanding long-term results. In recent decades, there have been significant improvements in minimally invasive surgery (MIS), slowly igniting a revolution in the field of pediatric surgery. At the turn of the twenty-first century, the first case reports of laparoscopic duodenoduodenostomy in neonates was published. In the 20 years since, this method has been adopted somewhat slowly. This is likely due to high rates of conversion to open (up to 35%) and anastomotic complications (up to 33%) relative to open repair in the initial years, as well as the inherent complexity of performing delicate dissection and intracorporeal suturing in a limited working space.

In recent years, there has been increased focus on MIS, and the skills of pediatric surgeons, predominantly at tertiary care centers, and these skill sets have evolved significantly. Laparoscopic duodenoduodenostomy offers patients smaller incisions promoting earlier recovery and discharge; the lack of distal bowel manipulation may result in a shorter ileus and earlier initiation of oral feeding. Potential disadvantages of MIS repair include longer operative time (mean operative time 30 min longer) and increased difficulty assessing for lesions such as webs. A recent meta-analysis and systematic review demonstrate laparoscopic duodenoduodenostomy conversion to open and complication rates at 17% and 16%, respectively. Thus, MIS repair performed by those with sufficient laparoscopic skills provides comparable safety and efficacy to open repair without a significant difference in complication rates.

The use of endoscopy in the management of duodenal webs in infants was first reported in 1989 by T Okamatsu et al.; a high-frequency wave cutter was used to make a T-shaped incision in the duodenal web. Since then, the success of various endoscopic techniques and instruments have been reported; these include electrocautery, laser resection, insulated tip endo-knife, biliary needle knife, suction with high-frequency wave snare, papillotome, sphincterotome, and balloon dilation. Given the risk of bowel perforation and bleeding with the use of electrocautery and/or laser, these are not recommended.

Membranectomy via balloon dilation and division using an insulated tip endo-knife are documented successful techniques. However, the ideal approach involves the usage of a double-channel endoscope, passing a guidewire through the membrane fenestration, inflation of the balloon distally, retraction of the web toward the scope, and creation of a radial incision in the web using an insulated tip endo-knife. Care must be taken to avoid incising medially as the pancreaticobiliary channels are found here; endoscopic ultrasound is often used to identify and avoid these structures. A radial incision has shown to be less likely to form strictures than circumferential incisions, which are not usually necessary to relieve the obstruction. An endoscopic approach to duodenal webs spares the infant from laparotomy, decreases the risk of postoperative intra-abdominal adhesions, and allows for faster recovery and shorter hospitalization. While therapeutic endoscopy is the preferred method for incomplete duodenal webs discovered later in life, reports of endoscopic treatment in infants are far more limited. When deciding to manage infants endoscopically, the technical ability of the endoscopist and the availability of a pediatric surgeon must be considered.

## Study Questions

1) Regarding infants with duodenal atresia all the following statements are true *except*?

   A. The best option for surgical correction is a gastrojejunostomy.
   B. Prenatal ultrasound can detect duodenal atresia.
   C. There is an increased incidence of Trisomy 21.
   D. It may be seen in combination with annular pancreas or malrotation.

   Answer to Question 1: A—The ideal procedure to correct duodenal atresia or stenosis is to bypass the obstruction by performing a duodenoduodenostomy. If performing a duodenoduodenostomy is technically challenging due to the location of obstruction or patient-specific factors, the next best choice is a duodenojejunostomy. Gastrojejunostomy should not be used due to the potential for future complications such as marginal ulcers or blind loop syndrome. Duodenal atresia may be diagnosed prenatally when the "double-bubble" sign is seen on ultrasound. Trisomy 21 is present in up to half of infants with duodenal atresia or stenosis. Annular pancreas or malrotation may concomitantly occur in these patients.

2) The pathogenesis of congenital duodenal atresia is most likely the result of:

   A. *In utero* vascular accidents

B. Failure of luminal recanalization
C. A & B
D. None of the above

Answer to Question 2: B—Whereas intestinal atresia distal to the duodenum are the consequence of intrauterine vascular accidents, atresia of the duodenum is unique. During the sixth week, the gut epithelium rapidly proliferates causing complete occlusion of the intestinal lumen. Over the course of the eight through tenth weeks, epithelial apoptosis occurs, gradually recanalizing and re-establishing continuity of the now epithelium-lined intestinal lumen. Failure to recanalize leads to duodenal atresia.

3) You are evaluating a 1-day-old baby boy with bilious emesis; he has Trisomy 21 and epigastric fullness. Abdominal plain film shows a "double-bubble" sign. A week later, after appropriate resuscitation, he is taken to the operating room and a windsock duodenal atresia deformity is found to be the cause of obstruction. The best procedure for repair is:

A. De-rotation of the intestines
B. Gastrojejunostomy
C. Lateral tapering duodenoplasty
D. Diamond duodenoduodenostomy with excision of the obstructing band

Answer to Question 3: D—A windsock deformity of the duodenum can be repaired with careful excision of the obstructing band, a duodenoduodenostomy, or both. De-rotation of the intestines is used for malrotation. Gastrojejunostomy should not be used due to the increased likelihood of future complications. A lateral tapering duodenoplasty can be performed at the time of initial operation to facilitate peristalsis or later in cases where persistent duodenal dilation prevents effective peristalsis.

**Illustrations**

1) Illustration 1: Types of Duodenal Atresia
2) Illustration 2: Plain Abdominal Radiograph showing classic "double-bubble" sign.
3) Illustration 3: Creation of a diamond-shaped anastomosis.

## Further Reading

Al-Salem AH. Congenital duodenal obstruction. Atlas of pediatric surgery. Springer; 2020:433–443: chap 48.

Applebaum H, Sydorak R. Duodenal atresia and stenosis-annular pancreas. Chapter 81. In: Coran AG, Caldamone A, Adzick NS, Krummel TM, Laberge JM, Shamberger R, editors. Pediatric surgery. 7th ed. Philadelphia: Elsevier Pub; 2012. p. 1051–8.

Bax NM, Ure BM, van der Zee DC, van Tuijl I. Laparoscopic duodenoduodenostomy for duodenal atresia. Surg Endosc. 2001 Feb;15(2):217.

Coppola C, Kennedy A Jr, Scorpio R, editors. Pediatric surgery: diagnosis and treatment. Springer; 2014.

Escobar MA, Ladd AP, Grosfeld JL, et al. Duodenal atresia and stenosis: long-term follow-up over 30 years. J Pediatr Surg. 2004;39(6):867–71.

Grosfeld JL, Rescorla FJ. Duodenal atresia and stenosis. In: Gray SW, Skandalakis JE, editors. Embryology for surgeons: the embryological basis for the treatment of congenital defects. Philadelphia: WB Saunders; 1986. p. 33–8.

Juliet MC, Clark B, et al. The double bubble sign: duodenal atresia and associated genetic etiologies. Fetal Diagn Ther. 2020;47(2):98–103.

Kimura K, Loening-Baucke V. Bilious vomiting in the newborn: rapid diagnosis of intestinal obstruction. Am Fam Physician. 2000;61(9):2791–8.

Kimura K, Mukohara N, Nishijima E, Muraji T, et al. Diamond-shaped anastomosis for duodenal atresia: an experience with 44 patients over 15 years. J Pediatr Surg. 1990;25:977–9.

Lamoshi AY, Abdulhai S, Ponsky TA. Duodenal obstruction in newborns. Springer. 2019:499–500.

Mentessidou A, Saxena AK. Laparoscopic repair of duodenal atresia: systematic review and meta-analysis. World J Surg. 2017;41:2178–84.

Ogle S, Nichol P, Ostlie DJ. Duodenal and intestinal atresia and stenosis. In: Holcomb and Ashcraft's pediatric surgery. 7th ed. Philadelphia: Saunders/Elsevier; 2020.

Okamatsu T, Arai K, Yatsuzuka M, Ishikawa M, et al. Endoscopic membranectomy for congenital duodenal stenosis in an infant. J Pediatr Surg. 1989;24(4):367–8.

Rothenberg SS. Laparoscopic duodenoduodenostomy for duodenal obstruction in infants and children. J Pediatr Surg. 2002;37(7):1088–9.

Sweed Y, Vachyan A. Pediatric surgery. In: Puri P, Hollwarth M, eds. Pediatric surgery. 2nd ed. Springer 2019:191–200: chap 25.

Vinycomb T, Browning A, Jones MLM, Hutson JM, King SK, Teague WJ. Quality of life outcomes in children born with duodenal atresia. J Pediatr Surg. 2020;55(10):2111–4.

Sweed Y, Yulevich A. Duodenal obstruction. In: Puri P, editor. Pediatric surgery. Berlin: Springer; 2019. https://doi.org/10.1007/978-3-642-38482-0_63-2.

# Intestinal Atresia

**Alysia A. Agnoni and Christopher P. Coppola**

**Key Points**
1. Jejuno-ileal atresias (JIA) most likely occur due to vascular or ischemic insult to the mesenteric blood supply in utero.
2. Small bowel atresias are classified into four different categories and each occurs in relatively equal frequency, aside from Type IIIB, also known as the apple-peel atresia, which is the least common.
3. Short bowel syndrome, a long-term complication of intestinal atresia, is more commonly associated with Type IIIb or IV JIA.

1. Background:
   a. Intestinal atresia is a congenital discontinuity of the gut which results in obstruction and vomiting in neonates. Jejuno-ileal atresias (JIA) are most commonly categorized together as one entity and can occur as a single obstruction or as several lesions. Continuity of the bowel can usually be restored, but there is a risk of short gut syndrome and inability to survive on enteral feeding alone. Duodenal atresia is thought to result from a different pathophysiology and is discussed separately.

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2. Pathophysiology:
    a. Epidemiology
        i. Incidence: Intestinal atresia occurs in one in every 5000–10,000 live births with equal distribution between males and females. Malrotation can also be present in up to 25% of cases. Gastroschisis and cystic fibrosis are also associated with increased risk of a JIA. Maternal risk factors include smoking or cocaine use during pregnancy.
        ii. Etiology
            1. Intestinal atresia is thought to be the result of mesenteric vascular occlusion during gestation. This could be due to strangulation via an intestinal hernia, intussusception, volvulus, or thromboembolism. The segments of intestine without blood flow during development become atretic. In cases when vascular compromise is brief or incomplete, the result may be intestinal stenosis instead.
            2. Hereditary multiple intestinal atresia (HMIA) is very rare autosomal recessive disorder that is considered fatal.
    b. Location: Any part of the gastrointestinal tract can be affected. Some studies suggest that the terminal ileum is the most common site of atresia, but not all studies support this. Atresia of the colon is rare.
    c. Classification: (Fig. 1).
        i. Type I: Atretic bowel has no lumen, but muscular wall is intact.
        ii. Type II: The atretic segment is a fibrous cord, with no muscular wall.
        iii. Type III: There is absence of the bowel and a wedge-shaped gap in the mesentery.
            1. Type IIIa: Simple defect of limited length.
            2. Type IIIb: This is the "Christmas tree" or "Apple peel" deformity where the bowel ends blindly shortly after the ligament of Treitz. The only distal bowel present is a stenotic spiral wrapped around the ileocecal artery like the coiled peel from an apple. The base of the coil is wider than the tip, giving the cone-shaped appearance of a Christmas tree (Fig. 2).
        iv. Type IV: Multiple segments of intestinal atresia.
3. Clinical features:
    a. Infants will present shortly after birth with abdominal distention and bilious vomiting. If the atresia is distal, it may take several feeds before the gut fills enough for there to be emesis. Perforation can occur and patients will have signs of sepsis and shock. Atresia of the intestine can happen in mid-gestation after the infant has swallowed amniotic fluid containing epithelial cells and lanugo. Therefore, passage of meconium after delivery does not necessarily rule out an intestinal atresia.

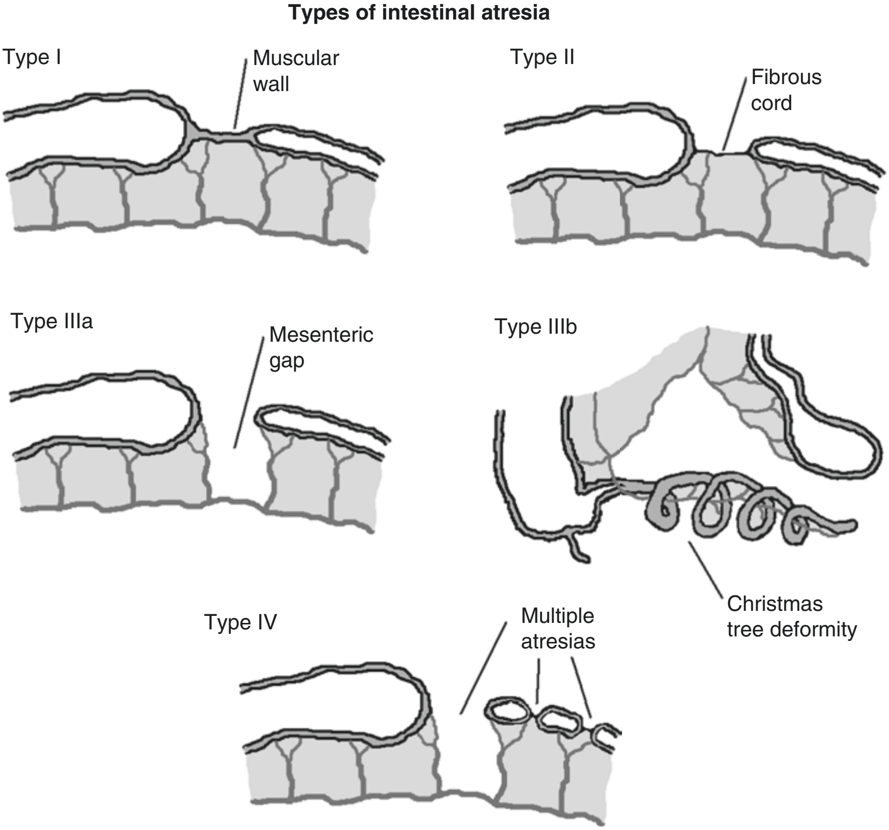
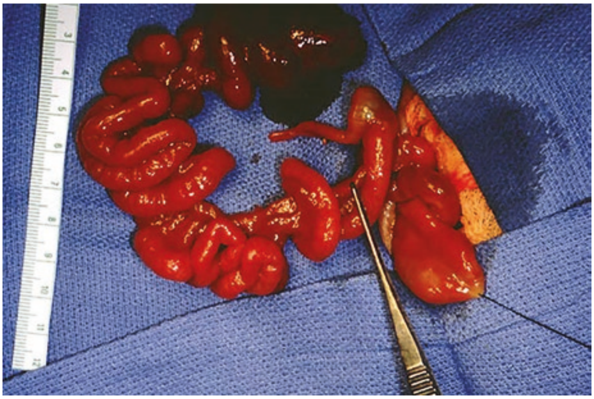

Types of intestinal atresia
Type I
Muscular wall
Type II
Fibrous cord
Type IIIa
Mesenteric gap
Type IIIb
Christmas tree deformity
Type IV
Multiple atresias

**Fig. 1** Types of intestinal atresia. In Type I, there is a muscular wall without lumen connecting the two ends of atretic bowel. In Type II, they are connected by a fibrous cord. In Type IIIa, there is a wedge-shaped gap in the mesentery between the two ends. Type IIIb is a severe form of the disease in which the only distal bowel present is spiraled around a branch of the ileocecal artery. Type IV is multiple atresias. (Source: Christopher Coppola)

**Fig. 2** Intestinal atresia Type IIIb: "Christmas tree" type deformity. The loop of bowel with the black suture is the distended blind end of the distal duodenum, the bowel loops next to the ruler are the unused small diameter distal small bowel coiled around a solitary ileocecal artery. (Source: Christopher Coppola)



4. Diagnosis:
    a. Prenatal sonography may reveal polyhydramnios. It may also reveal distended or echogenic loops of bowel.
    b. Abdominal X-ray can be diagnostic and will reveal distended proximal loops of bowel without distal gas. In some cases, there will be a "triple bubble" sign showing a dilated stomach, duodenum, and proximal jejunal segment. If perforation has occurred, there may be pneumoperitoneum.
    c. Fluoroscopy with contrast can reveal the site of obstruction. When an infant presents with bilious vomiting, it is often a dilemma deciding between upper gastrointestinal series and contrast enema. As the most important lesion to diagnose and treat rapidly is malrotation with midgut volvulus, it is usually better to obtain upper gastrointestinal series first to rule out this condition. Contrast enema can be obtained after upper gastrointestinal series, but it may be limited in interpretation due to contrast in proximal gut.
5. Initial management:
    a. Patients should be made NPO and a nasogastric tube is placed to decompress the stomach.
    b. Intravenous access is established, patients are given resuscitative and replacement fluid administration, and any electrolyte abnormalities are corrected.
    c. Broad spectrum antibiotics are considered in case of perforation or evidence of infection.
6. Surgical treatment:
    a. Intestinal atresia is repaired with laparotomy and re-anastomosis of the two blind-ending loops of bowel. A laparoscopic-assisted approach can be considered but is generally felt to be more challenging and somewhat unnecessary given that neonatal bowel can be exteriorized through a very small incision.
    b. Transverse incision superior to the umbilicus will give the best view of the gut from duodenum to rectum.
    c. Bowel is run to identify the location of atresia, evaluate for multiple atresias, and determine if any other intra-abdominal abnormality is present.
    d. The bowel is anastomosed using an end (of proximal dilated gut) to back (antimesenteric surface of narrow caliber distal gut). The most dependent portion of proximal gut is used to encourage emptying. Sometimes tapering enteroplasty is needed for extremely dilated gut, but this can be performed at a future operation only if needed. The distal gut is much smaller in diameter, so an antimesenteric slit is made until the aperture matches the diameter of the proximal gut. Prior to closure of anastomosis, a feeding tube is threaded into distal gut to irrigate with saline to prove that there are no additional obstructions downstream.
    e. One may consider a diverting ostomy in the setting of significant size discrepancy between the dilated proximal portion and distal end. Stoma can also be useful in cases of multiple atresias or intestinal perforation.

   f. There is often a long delay to oral feeding after repair of intestinal atresia, so consideration is made for a central line for parenteral access.
   g. A nasogastric tube should be placed for postoperative decompression of the stomach while waiting for bowel function.

7. Complications:

   a. As with all operative procedures, post-op wound infections can occur. Anastomotic leak or stenosis and adhesive bowel obstructions are other risks. There may also be feeding intolerance or intestinal failure, especially if there was significant size discrepancy between proximal and distal ends, intestinal perforation, or short bowel syndrome.
   b. Short bowel syndrome:

      i. More commonly associated with Type IIIb or IV atresias.
      ii. Infants may require prolonged total parental nutrition (TPN) via a central line until intestinal function is re-established.

8. Prognosis:

   a. Overall survival is greater than 90%. Infants with more distal lesions have a better prognosis, whereas poorer outcomes have been associated with Type IIIb or IV atresia, prematurity, low birth weight (<2 kg), and other associated congenital anomalies. In infants with short bowel syndrome, bowel lengthening procedures or intestinal transplant can be considered.

## Additional Notes

Colonic atresia is the rarest form of intestinal atresia and is associated with Hirschsprung's disease and gastroschisis. The etiology is suspected to be similar to small bowel atresia with there being an in utero vascular event leading to atresia. The right colon is more often affected. A primary repair with excision and anastomosis is possible, especially with a right-sided defect. Left-sided colonic atresia may require a staged repair. It is wise to also perform a distal rectal biopsy to rule out aganglionosis (Hirschsprung's disease) prior to anastomosis.

## Study Questions

1. Which type of intestinal atresia is associated with short bowel syndrome and carries the poorest prognosis?

   a. Type I
   b. Type II
   c. Type IIIa
   d. Type IIIb

Answer: (d) Type IIIb. Type IIIb intestinal atresia, also known as the “Christmas tree” or Apple peel” deformity. This is a wedge defect of the small bowel mesentery that extends all the way from a blind ending duodenum to the ileocecal artery. The stenotic distal bowel is often coiled around this solitary ileocecal artery with a wider base, giving the appearance of Christmas tree around a central trunk. The lack of proximal bowel and the underdeveloped stenotic nature of the unused distal bowel often lead to short bowel syndrome and a worse prognosis.

2. Which of the following is NOT associated with increased risk of intestinal atresia?
   a. Gastroschisis
   b. Malrotation
   c. Omphalocele
   d. Cystic Fibrosis

Answer: (c) Omphalocele. Intestinal atresia can be associated with gastroschisis and cystic fibrosis. Often in intestinal atresia, the abnormal and incomplete anatomy of the small bowel is associated with malrotation and abnormal or absent fixation of the bowel. Colonic atresia can be associated with Hirschsprung’s disease or gastroschisis. There is not an association between intestinal atresia and omphalocele.

3. Which incision will provide the best view of intestinal anatomy to evaluate for an intestinal atresia (duodenum to rectum)?
   a. Supraumbilical transverse incision
   b. Infraumbilical transverse incision
   c. Circum-umbilical incision
   d. Laparoscopic-assisted

Answer: (a) Supraumbilical transverse incision. Most abdominal operative interventions in neonates are best achieved through a supraumbilical transverse incision. Infants and children with a relatively shorter or wider torso will have more room for access achieved with a transverse incision instead of a midline laparotomy. Other benefits of a transverse incision are more cosmetic healing because the incision is in Langer’s lines and less impairment of spontaneous ventilation because the incision does not come as close to the chest. For intestinal atresia, usually only a short incision is needed because the small bowel can be eviscerated through a small incision to perform running of the bowel and anastomosis to repair the atresia. An infraumbilical incision is less useful because eviscerating the small bowel through a lower incision is more difficult because it is further from the root of the superior mesenteric artery. A circum-umbilical incision is not typically used. The laparoscopic approach is not ideal because visualization is difficult in the small abdominal cavity of a neonate, and it is difficult to run the entire bowel to determine if there are multiple atresias.

## Further Reading

Adams SD, Stanton MP. Malrotation and intestinal atresias. Early Hum Dev. 2014;90:921–5. https://doi.org/10.1016/j.earlhumdev.2014.09.017.

Piper HG, Alesbury J, Waterford SD, et al. Intestinal atresias: factors affecting clinical outcomes. J Pediatr Surg. 2008;43:1244–8. https://doi.org/10.1016/j.jpedsurg.2007.09.053.

Bilodeau A, Prasil P, Cloutier R, et al. Hereditary multiple intestinal atresia: thirty years later. J Pediatr Surg. 2004;39:726–30.

Morris G, Kennedy A, Cochran W. Small bowel congenital anomalies: a review and update. Curr Gastroenterol Rep. 2016;18:16. https://doi.org/10.1007/s11894-016-0490-4.

Osuchukwu OO, Rentea RM. Ileal atresia. In: StatPearls [Internet]. Treasure Island: StatPearls Publishing; 2020.

# Short Bowel Syndrome and Intestinal Lengthening Procedures

**Pooja S. Salvi and Robert A. Cowles**

**Key Points**

1. Following small bowel resection, the intestine in patients with short bowel syndrome (SBS) undergoes compensatory adaptive changes to increase functional nutrient and water absorption. Loss of the distal ileum is associated with least favorable adaptation, owing to the presence of specialized epithelial cells in the ileum that are critical for fat absorption, and which secrete hormones involved in intestinal transit and mucosal growth.
2. Intestinal loss and dependence on parenteral nutrition have significant physiologic consequences for patients with SBS. Multidisciplinary intestinal rehabilitation programs coordinate medical and surgical interventions for these patients, to optimize intestinal adaptation, prevent complications of SBS, and accelerate weaning from parenteral nutrition.
3. Small intestinal lengthening procedures and intestinal transplant are surgical interventions that can improve outcomes in patients with SBS. Intestinal lengthening procedures can increase bowel function and surface area, to aid in weaning from parenteral nutrition, whereas intestinal transplant can be considered in patients with parenteral nutrition failure.

1. Background

Pediatric short bowel syndrome (SBS) is a unique surgical problem, in that the etiology is most often surgical, whereas the treatment involves a truly multi-

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disciplinary approach in which surgery is rarely curative. Nonetheless, knowledge of the causes and pathophysiology of this disorder is critical for the pediatric surgeon, in order to facilitate timely intervention that can prevent prolonged patient morbidity.

Starvation from anatomic loss or functional failure of the gastrointestinal (GI) tract has long been a challenge encountered by pediatric surgeons caring for infants. In the 1960s, the advent of parenteral nutrition (PN) introduced the possibility of survival and growth in these patients and opened the door for surgeons to operate on infants with congenital and acquired conditions of the intestine who otherwise would have had no chance of survival.

Pediatric intestinal failure (IF) is defined as the inability to absorb sufficient nutrients and water to support a child's growth and hydration. Short bowel syndrome (SBS) is a chronic or protracted form of IF, characterized by insufficient nutrient absorption after massive small bowel loss, and is the most common cause of IF in the pediatric population. Other causes of IF include GI conditions resulting in malabsorption from mucosal enteropathy and disorders of intestinal motility.

2. Epidemiology: causes and incidence

In most cases, pediatric SBS is caused by congenital or acquired conditions of the newborn that result in massive bowel resection or functional bowel loss. Table 1 lists the most common etiologies. Necrotizing enterocolitis (NEC) is the leading cause of pediatric SBS, with highest incidence in premature infants. Acute management of NEC aims to control abdominal sepsis, sometimes requiring laparotomy and resection of ischemic bowel leading to SBS. Efforts to prevent prematurity, which is the most important risk factor for NEC, and to identify the underlying etiology of this disease process have not been successful in decreasing the incidence. Simultaneous advances in neonatal intensive care have increased survival of infants with NEC and thus resulted in more children requiring management after massive bowel resection.

Indeed, there have been reports that the prevalence of pediatric SBS may be rising based on the increased prevalence of common underlying disorders. Specifically, the mean incidence of NEC has increased from 3–11% between 1997 and 2000 to 5–15% between 2003 and 2007. Gastroschisis, another significant cause of SBS, has increased in incidence by 30% from 1995–2005 to 2006–2012. As a congenital disorder of the abdominal wall, gastroschisis can

**Table 1** Common causes of pediatric short bowel syndrome

| |
|---|
| Necrotizing enterocolitis |
| Gastroschisis |
| Intestinal atresia |
| Midgut volvulus |
| Total or subtotal intestinal aganglionosis |
| Vascular thrombosis |
| Trauma |
| Inflammatory bowel disease |

require bowel resection due to intestinal injury or ischemia. Prolonged intestinal dysmotility can also affect newborns with gastroschisis, further contributing to the challenge of weaning PN, even in the absence of massive bowel resection.

In general, the overall incidence and prevalence of pediatric SBS are difficult to measure. The biggest reason being SBS is a functional state defined by need for PN, rather than an anatomic one. As such, diagnostic criteria for SBS based on duration of PN-dependence are not uniform across institutions or even providers. Second, the provision of PN can occur in a number of different inpatient and outpatient venues, making it difficult to track patients over time. In one study, SBS defined as the need for PN for more than 42 days after bowel resection or a residual small bowel length of less than 25% of that expected for gestational age yielded an incidence of 24.5 cases per 100,000 live births.

3. Pathophysiology of short bowel syndrome

To understand the pathophysiologic processes that contribute to PN-dependence in SBS, it is important to first understand normal intestinal physiology. In the intact GI tract, the small intestine is the primary site of nutrient absorption. Intestinal villi, the finger-like projections of the small intestinal mucosa, are lined with enterocytes that have the capacity to absorb nutrients from luminal contents. The villus configuration of the mucosa contributes to the enormous absorptive surface area of the small intestine, which is greatest in the proximal small bowel and least in the distal ileum. This proximal-to-distal gradient reflects the concentration of nutrients and macromolecules present in the different regions of the intestine. The majority of macronutrients undergo mechanical and chemical digestion in the stomach and proximal intestine, and subsequently undergo more thorough digestion by intestinal brush border enzymes, followed by absorption in the small intestine. Additionally, the small intestine is an essential site of fluid and electrolyte absorption, responsible for absorbing 80% of the fluid that passes through the GI tract.

In pediatric SBS, disruption of the GI tract from small bowel resection can result in one of three anatomic configurations of the remnant intestine: (1) small intestinal resection with a small bowel (typically jejunoileal) anastomosis, (2) small bowel and partial colon resection with an enterocolonic anastomosis, or (3) small bowel resection with a jejunostomy (Fig. 1). Each of these configurations has a unique pathophysiology, with implications for disease severity and management.

A. Small bowel resection with jejunoileal anastomosis

Patients who undergo resection of only the small intestine with subsequent jejunoileal anastomosis rarely experience major changes in nutrient or water absorption, though this depends to some degree on the length of intestine removed. The remnant small intestine and colon have the capacity to undergo compensatory histologic and functional changes over time. After bowel resection, the small intestinal epithelium grows longer intestinal villi and exhibits enhanced nutrient absorptive capacity, and the colon is capable of providing energy by way of microbial production of short-chain fatty

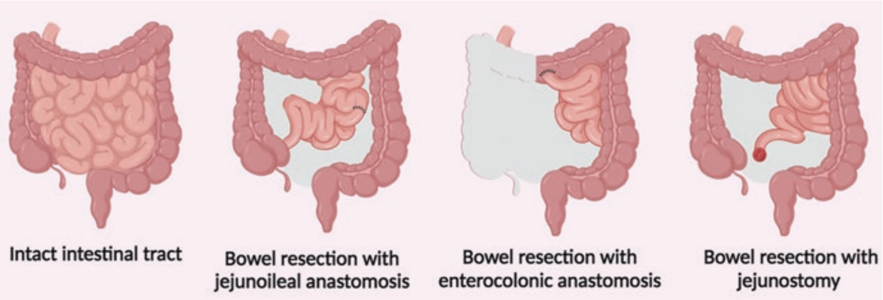
Intact intestinal tract
Bowel resection with jejunoileal anastomosis
Bowel resection with enterocolonic anastomosis
Bowel resection with jejunostomy

**Fig. 1** Anatomic configurations of the intestinal tract in patients with short bowel syndrome after small bowel resection. The most favorable post-bowel resection anatomy is achieved with an intact ileocecal valve and intestinal continuity via a small bowel (e.g., jejunoileal) anastomosis. Resection of the distal ileum and ascending colon with an enterocolonic anastomosis is less favorable, due to loss of specialized epithelial cells in the distal small intestine. An end jejunostomy, in which the distal small intestine and colon are resected or are not contiguous with the proximal intestinal tract, is the least favorable anatomy due to both loss of specialized ileal mucosa and fluid losses from the stoma. (Source: Pooja S. Salvi and Robert A. Cowles)

acids (SCFA), which serve as fuel for colonocytes. Additionally, the colon has a large reserve capacity for fluid absorption, which is utilized under conditions of small bowel loss.

B. Distal small bowel resection with enterocolonic anastomosis

Patients who undergo ileal and ileocolonic resections fare worse than patients undergoing more proximal bowel resections. This is in part due to the relatively diminished capacity for functional adaptation in the proximal small intestine, relative to the ileum. Additionally, ileal resection can result in loss of bile salt resorption and aberrations in GI hormone secretion since bile salt transporters and specific hormone-secreting cells are specifically expressed in the mucosa of this intestinal region. Consequently, insufficient enterohepatic bile circulation can result in fat malabsorption and steatorrhea. Loss of the ileocecal valve at the distal end of the small bowel may also impair GI function and the ability to wean a child from PN.

C. Jejunostomy

As expected, patients who require a jejunostomy after bowel resection have worse outcomes than the previous two anatomic configurations, owing to the loss of functional adaptation and physiologic reserve exhibited by the distal small intestine and colon. In patients with less than 100 cm of small bowel proximal to the jejunostomy, dehydration is a major concern, due to net fluid losses from the ostomy that cannot be adequately resorbed. Furthermore, changes in intestinal hormone secretion following jejunostomy result in accelerated gastric emptying and intestinal transit. In addition to potentially worsening high-volume stomal losses, acceleration of intestinal transit allows for less contact time between luminal contents and the mucosa for absorption. Refeeding of a distal mucous fistula can help achieve increased enteral nutrition and fluid absorption as well as preventing villous atrophy of the defunctionalized segment.

D. Extra-intestinal consequences of SBS

Altered intestinal anatomy and physiology in children with SBS can lead to pathology outside the GI tract, often resulting from abnormal nutrient absorption. Metabolic bone disease is common in this population, owing to increased calcium excretion, toxicity from PN, and overall malnutrition. Additionally, bile salt deficiency due to loss of bile salt transporters in the ileum predisposes patients to sequelae of inadequate fat absorption including formation of calcium oxalate kidney stones and gallstones. In SBS, fatty acids present in the distal intestine due to fat malabsorption preferentially bind intraluminal calcium, which under normal conditions is bound by oxalate. As a result, oxalate is freely absorbed into the bloodstream in the colon and subsequently undergoes renal excretion, thus increasing the risk of calcium oxalate nephrolithiasis. Similarly, patients with diminished bile acids are unable to effectively solubilize cholesterol, which increases the likelihood of cholesterol gallstone formation due to supersaturation of cholesterol in bile. Other physiologic alterations in SBS can also increase risk for cholelithiasis, including altered bilirubin metabolism and altered GI hormone secretion leading to reduced gallbladder contractility.

Importantly, loss of specialized cells in the distal intestine that produce regulatory GI hormones, such as cholecystokinin and secretin, can disrupt hormonal feedback mechanisms that modulate GI motility and intestinal gastrin production. Hypergastrinemia and associated hyperacidity may necessitate treatment with proton pump inhibitors or histamine receptor blockers, though these signs are typically transient, lasting for weeks to months after intestinal resection.

GI dysmotility after intestinal surgery and achlorhydria from pharmacologic acid suppression contribute to small intestinal bacterial overgrowth (SIBO) in children with SBS. This condition, characterized by changes in the normal enteric microbiome, can lead to mucosal damage from excessive epithelial inflammation and diminished mucosal nutrient absorption. D-lactic acidosis is a specific metabolic abnormality associated with intestinal bacterial dysbiosis that occurs when the presence of abnormal gut flora overwhelms normal bacterial metabolism of D-lactate, a product of carbohydrate metabolism, in the colon. Accumulation of D-lactate in the bloodstream can manifest in children as neurologic symptoms ranging from lethargy and poor school performance to coma.

Diagnosis of SIBO in SBS is challenging due to the nonspecific constellation of symptoms and the difficulty of accurately and non-invasively assessing the bacterial composition of the small intestine. Direct culture of small bowel fluid and hydrogen breath tests are among the currently utilized methods to diagnose SIBO, though variation in the microbial species present in individual patients can obscure interpretation of findings.

E. Enteral autonomy

As mentioned previously, proof that the metabolic and growth demands of children could be sustained with parenteral nutrition in the 1960s opened up possibilities for surgeons to intervene on conditions that would have previously left a child with a non-survivable amount of intestine. Though PN formulations and delivery have improved substantially over the past 50 years, the goal of treatment in pediatric SBS remains achieving enteral autonomy or weaning from PN.

A number of anatomic and physiologic factors contribute to the likelihood of achieving enteral autonomy. Residual bowel length relative to expected bowel length is the primary determinant of outcome. However, since potential for gut growth is closely correlated to an infant's stage of development, it is important to consider a child's gestational age when calculating expected bowel length. At the beginning of the third trimester, infant small bowel is approximately 125 cm in length, whereas a term infant has approximately 200 cm of small bowel and 40–50 cm of colon. To account for gestational age, it is common practice to present residual small bowel as a percentage of the total bowel length expected for a patient based on their age. Multiple studies have demonstrated that small bowel length greater than 10% of expected is a predictor for enteral autonomy and survival in infants with SBS.

Location of resected bowel is another significant predictor of weaning from PN. Preservation of the ileocecal valve is a predictor of enteral autonomy. Though this was previously attributed to possible transit-slowing mechanisms of the valve, this advantage is more likely related to properties of the ileal mucosa itself. For example, specialized function and hormone secretion from the ileum, such as secretion of glucagon-like peptides 1 and 2, may enhance intestinal adaptation, and the distal intestine exhibits a greater capacity for adaptive growth than proximal bowel regions.

## 4. Treatment

### A. Multidisciplinary intestinal rehabilitation programs

Intestinal adaptation, including the functional absorptive changes in the remnant intestine, begins shortly after bowel resection and is typically complete within 24–60 months. Intestinal rehabilitation (IR) refers to the supportive medical and surgical interventions that can maximize this adaptive response and thus accelerate an infant's time to enteral autonomy. IR is a multidisciplinary approach that includes both proactive and prophylactic management strategies and is the mainstay for pediatric SBS treatment. Optimization of enteral and parenteral nutrition formulations is a central element of IR, in order to maintain a patient's growth, while promoting intestinal adaptation. Medical management is further necessary to prevent and promptly address highly morbid complications of PN, such as central-line-associated bloodstream infections (CLABSI) and PN-associated liver disease.

PN-associated liver disease (PNALD) is a strong predictor of mortality in children with SBS, and protocols to aggressively prevent progression of

PNALD have been successful in improving patient survival. The pathogenesis of PNALD is likely multifactorial. Decreased enteral nutrient intake can compromise the intestinal epithelial barrier and allow passage of enteric bacteria into the hepatic circulation, where subsequent endotoxin release, inflammatory cytokine production, and sepsis result in liver damage. Additionally, there is evidence that PN itself is hepatotoxic. Intravenous lipid emulsions (ILE) in PN formulations contain proinflammatory components, such as omega-6 fatty acids, which induce hepatocyte apoptosis and cause cholestasis. Furthermore, there is evidence that ILE are associated with loss of specialized biliary transport proteins, which are responsible for transporting components of bile from hepatocytes into bile ducts. Downregulation of these transport proteins is associated with inflammation and fibrosis of the biliary system and liver parenchyma.

Previously, intravenous lipid restriction was considered as an approach to minimize PNALD. Though successful in slowing the progression to PNALD, which is defined as serum conjugated bilirubin greater than 2 mg/dL, lipid restriction predisposed children to complications associated with essential fatty acid deficiency. As such, more recent approaches to addressing the deleterious effects of ILE involve substitution of traditional soybean-based lipid emulsions, which appear to be particularly hepatotoxic, with novel fish oil-based emulsions or mixed (composite) lipid sources. Fish oil-based emulsions lack phytosterols, a plant-derived cholesterol-like molecule found in soybean-derived lipids that have been associated with progression of PN-associated cholestasis and PNALD.

Current consensus guidelines on pediatric parenteral nutrition recommend the use of fish oil-containing lipid emulsions due to the advantages it demonstrates over soybean-derived lipids, namely, reduced risk of cholestasis and reduced liver inflammation, while maintaining adequate provision of essential fatty acids for neurodevelopment and metabolism. This includes composite lipid emulsions containing soybean oil, medium chain fatty acids, olive oil, and fish oil (SMOF). In general, SMOF should be considered as first-line treatment in children with SBS who have cholestasis or other signs of PNALD.

Treatment of other comorbidities of SBS, such as kidney stones and small intestinal bacterial overgrowth, are components of IR that minimize insults to quality of life, morbidity, and barriers to enteral autonomy. Longitudinal studies of SBS patients have demonstrated that integration into a multidisciplinary IR program was associated with increased survival, although not with increased attainment of enteral autonomy.

B. Surgical intervention

The pediatric surgeon is an essential member of the multidisciplinary IR team. Surgical interventions that can accelerate intestinal adaptation and weaning from PN include establishment of enteral feeding access (e.g., gastrostomy tube placement), closure of enterostomies to minimize fluid losses, establishment of central venous access for infusion of PN, and intestinal lengthening procedures.

Two surgical procedures to increase function and/or surface area of the remnant small intestine in SBS are most often considered. In 1980, Bianchi described the longitudinal intestinal lengthening and tapering (LILT) procedure. Although this approach does not increase functional absorptive surface area, it has been demonstrated to improve intestinal transit by correcting the gross intestinal dilation that can occur after bowel resection. In 2003, the serial transverse enteroplasty (STEP) procedure was introduced as an alternative to the LILT. As its name suggests, the STEP procedure involves making a series of alternating transverse or angulated partial transections across a segment of dilated intestine, thus lengthening the effective intestinal channel and increasing functional absorptive surface area (Fig. 2).

Patient survival after the LILT procedure and STEP procedure is estimated to be 81% and 89%, respectively. Infants with greater than 50 cm of remnant intestine and no signs of clinical jaundice at the time of surgery are most likely to survive. Comparison of outcomes between patients who underwent an intestinal lengthening procedure or did not is generally difficult to make, due to less severe illness in children who are candidates for surgical intervention.

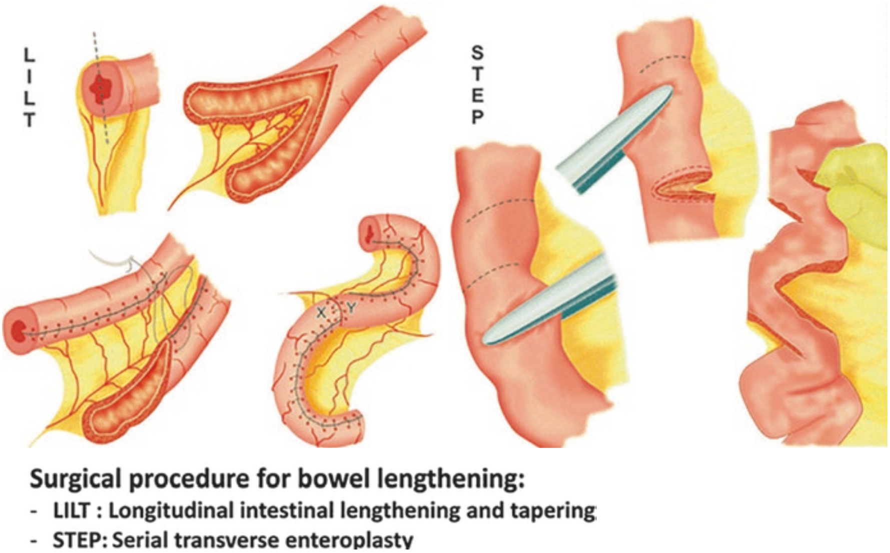
LILT
STEP
X
Y
Surgical procedure for bowel lengthening:
- LILT : Longitudinal intestinal lengthening and tapering
- STEP: Serial transverse enteroplasty

**Fig. 2** Intestinal lengthening procedures such as the longitudinal intestinal lengthening procedure (LILT) and the serial transverse enteroplasty procedure (STEP) are surgical interventions that can aid in optimizing intestinal adaptation after small bowel resection, by increasing intestinal function and absorptive surface area of the remnant small intestine. (Source: Goulet O, Lacaille F, Lambe C. Intestinal failure: etiologies and outcomes and decision-making between rehabilitation and transplantation. In: Dunn S, Horslen S, editors, Solid organ transplantation in infants and children. Organ and tissue transplantation. Springer, Cham, 2017. Used under RightsLink License No. 5193281062030, 20 Nov 2021)

An estimated 53% of children achieve enteral autonomy after an intestinal lengthening procedure, and the average time to enteral autonomy after the STEP procedure was 21 months. These figures are important to consider in selecting infants who may benefit from surgical intestinal lengthening. Specifically, patients should have adequate liver function, no evidence of portal hypertension, shortened bowel length, and sufficient physiologic reserve to survive long enough to achieve enteral autonomy. Furthermore, infants with short segments of highly dilated bowel may reap the benefit of improved intestinal motility after the LILT or STEP procedures.

A notable risk of both of these procedures is recurrent bowel dilation. In such cases, a repeat STEP or STEP after LILT can occasionally be performed. Other possible complications include staple line leak, obstruction, bleeding, and enteric fistula formation.

C. Intestinal transplant

In the late 1980s, multiple pioneering surgeons began having success in the field of visceral transplant. Goulet reported the first isolated small intestinal transplant with long-term survival in 1987, followed by the first successful combined liver–small intestinal transplant by Grant in 1989.

Small bowel transplant is currently offered to individuals with SBS who experience PN failure. In children, this includes failure to maintain growth, development, and hydration while receiving PN. PN failure also refers to the development of PN-related complications, such as recurrent catheter-related infections, thrombosis of two of the six major central venous access sites, and the progression of PN-associated liver disease. Multivisceral transplantation, which involves complete replacement of all organs in the abdominal cavity, may be indicated in the setting of porto-mesenteric thrombus or complex enteric fistula, as well as in the more common case of liver failure with portal hypertension. In children, combined liver–small intestinal transplant is performed most commonly (50%), followed by isolated small intestinal transplant (37%) and multivisceral transplant (13%).

With advances in immunosuppression, surgical technique, and monitoring for post-transplant rejection over the last two decades, survival following isolated intestinal transplant and multivisceral transplant has dramatically improved. In 2007, the 1- and 5-year graft survival rates for pediatric intestinal transplant were 74.2 and 483%. However, the potential morbidity of the operation and of long-term immunosuppression in children with poor physiologic reserve from GI malfunction cannot be understated. Organ rejection is a feared complication, which portends poor outcomes. In a report of 108 pediatric patients undergoing intestinal or combined liver–intestinal transplantation at a single institution, approximately 19% of children experienced severe rejection, among which one-third did not survive and 43% required graft removal and re-transplantation.

Following transplantation, IR remains a critical part of long-term recovery. The anatomic reconfiguration of the viscera, in addition to rejection episodes, ischemia-reperfusion, and altered lymphatic drainage can compromise tolerance of enteral feeding. Studies have shown that some degree of enteral malabsorption remains after intestinal transplant in children, but compensatory hyperphagia and increased caloric intake can allow for normal growth and development.

## Additional Notes

Pediatric SBS is the most common cause of pediatric intestinal failure, typically occurring after massive small bowel resection in the newborn period. With increases in neonatal intensive care and prenatal diagnosis, the incidence of SBS has increased and will likely continue to rise. Achieving complete enteral nutrition while maintaining adequate growth and hydration is the goal of treatment for all infants with SBS. Extent and location of bowel loss are significant predictors for whether a child will be able to achieve enteral autonomy, with resection of the ileum being least favorable. The use of multidisciplinary IR programs is essential to providing coordinated care and interventions that maximize intestinal adaptation following resection. Surgical intervention, including intestinal lengthening procedures, can increase intestinal surface area and function in select patients that develop dilated bowel. In children with PN failure, intestinal transplant can be considered, though graft rejection carries with it a poor prognosis.

## Study Questions

Question 1. A 6-week-old infant girl is being cared for in the neonatal intensive care unit. She was born at 31 weeks gestation, and developed necrotizing enterocolitis on day of life 7, requiring a laparotomy and resection of ischemic bowel. Which of the following statements is true?

a. Central venous access is not necessary in this patient.
b. Prematurity is not a risk factor for necrotizing enterocolitis.
c. Small intestinal transplant should be the next surgical intervention for this patient.
d. Bowel resection puts this patient at risk for developing gallstones and kidney stones.

Answer: (d). Bowel resection puts this patient at risk for developing gallstones and kidney stones. Bowel resection, particularly distal bowel resection, will result in loss of specialized cells of the ileum that contain bile salt transporters. Diminished bile acids can lead to reduced solubilization of cholesterol and formation of cholesterol stones in the gall bladder. Furthermore, fat malabsorption will result in calcium binding free fatty acids in the intestine, rather than binding oxalate. As a result, oxalate will be absorbed in the colon and undergo renal excretion, where it can cause oxalate nephrolithiasis.

Question 2. Which of the following anatomic configurations will be least favorable for subsequent functional intestinal adaptation and enteral nutrition tolerance?

a. Resection of the distal small intestine and colon with an enterocolonic anastomosis, leaving the child with 40% of expected bowel length for age.
b. Resection of the proximal jejunum with a small bowel anastomosis, leaving the child with 40% of expected bowel length for age.
c. Resection of multiple small proximal and distal small bowel segments with multiple small bowel anastomoses, leaving the child with 70% of expected bowel length for age.
d. Resection of the distal small intestine and colon and creation of an end jejunostomy, leaving the child with 40% of expected bowel length for age.

Correct answer: (d). Resection of the distal small intestine and colon and creation of an end jejunostomy, leaving the child with 40% of expected bowel length for age. (Is least favorable) The distal small intestine has greater adaptive capacity than the proximal intestine, and thus loss of the ileum is least favorable for functional intestinal adaptation. Additionally, creation of a jejunostomy can pose a challenge for hydration, since the absorptive capacity of the colon is lost.

Question 3. A 4-year-old boy with a history of gastroschisis and bowel resection in infancy presents to the emergency room with abdominal pain and fevers. He receives at home parenteral nutrition through a vascular access catheter. Which of the following statements is false?

a. Catheter-associated blood stream infections are a significant cause of morbidity and mortality in patients with SBS.
b. Patients with distal small bowel loss are predisposed to developing cholesterol gallstones, due to diminished bile salt absorption.
c. Hematuria in this patient would raise suspicion for uric acid stone nephrolithiasis.
d. Long-term parenteral nutrition can lead to cholestatic injury of the liver.

Correct answer: (c). Hematuria in this patient would raise suspicion for uric acid stone nephrolithiasis. (Is false) Patients with short bowel syndrome may be predisposed to formation of calcium oxalate kidney stones, not uric acid stone nephrolithiasis.

## Further Reading

Dudrick SJ, Wilmore DW, Vars HM, Rhoads JE. Can intravenous feeding as the sole means of nutrition support growth in the child and restore weight loss in an adult? an affirmative answer. Ann Surg. 1969;169(6):974.

Engelstad HJ, Barron L, Moen J, Wylie TN, et al. Remnant Small bowel length in pediatric short bowel syndrome and the correlation with intestinal dysbiosis and linear growth. J Am Coll Surg. 2018;227(4):439–49.

Fanaroff AA, Stoll BJ, Wright LL, Carlo WA, et al. Trends in neonatal morbidity and mortality for very low birthweight infants. Am J Obstet Gynecol. 2007;196(2):147.e1–8.

Filler RM, Eraklis AJ, Rubin VG, Das JB. Long-term total parenteral nutrition in infants. N Engl J Med. 1969;281(11):589–94.

Goulet O, Ruemmele F. Causes and management of intestinal failure in children. Gastroenterology. 2006;130(2):S16–28.

Goulet O, Sauvat F. Short bowel syndrome and intestinal transplantation in children. Curr Opin Clin Nutr Metab Care. 2006;9:304–13.

Jones AM, Isenburg J, Salemi JL, Arnold KE, et al. Increasing prevalence of gastroschisis--14 States, 1995–2012. MMWR Morb Mortal Wkly Rep. 2016;65(2):23–6.

Khan FA, Squires RH, Litman HJ, Balint J, et al. Predictors of enteral autonomy in children with intestinal failure: a multicenter cohort study. J Pediatr. 2015;167(1):29–34.e1.

Modi BP, Langer M, Ching YA, Valim C, et al. Improved survival in a multidisciplinary short bowel syndrome program. J Pediatr Surg. 2008;43(1):20–4.

Neelis E, de Koning B, Rings E, Wijnen R, et al. The gut microbiome in patients with intestinal failure: current evidence and implications for clinical practice. J Parenter Enter Nutr. 2019;43:194–205.

Sommovilla J, Zhou Y, Sun RC, Choi PM, et al. Small bowel resection induces long-term changes in the enteric microbiota of mice. J Gastrointestinal Surg Off J Soc Surg Alimentary Tract. 2015;19(1):56–64. discussion 64

Sommovilla J, Warner BW. Surgical options to enhance intestinal function in patients with short bowel syndrome. Curr Opin Pediatr. 2014;26(3):350.

Spencer AU, Neaga A, West B, Safran J. Pediatric short bowel syndrome: redefining predictors of success. Ann Surg. 2005;242(3):403.

Squires RH, Duggan C, Teitelbaum DH, Wales PW, et al. Natural history of pediatric intestinal failure: initial report from the pediatric intestinal failure consortium. J Pediatr. 2012;161(4):723–8.

Tappenden KA. Pathophysiology of short bowel syndrome: considerations of resected and residual anatomy. JPEN. J Parenter Enter Nutr. 2014;38(1):14S–22S.

Wales PW, De Silva N, Kim J, Lecce L, et al. Neonatal short bowel syndrome: population-based estimates of incidence and mortality rates. J Pediatr Surg. 2004;39(5):690–5.

# Hirschsprung Disease

**Michael J. Allshouse and Lindsay H. Devereux**

**Key Points**
1. Hirschsprung disease is characterized by the absence of ganglion cells in a portion or the entirety of the colon. In rare cases, it extends into the small intestine.
2. Definitive diagnosis of Hirschsprung disease is made by performing a rectal biopsy showing absence of ganglion cells.
3. Surgical treatment of Hirschsprung disease involves resection of the aganglionic segment with pull-through of ganglion containing bowel to the anus.
4. Hirschsprung associated enterocolitis is a dangerous and potentially life-threatening complication of and must be recognized early and treated aggressively. It can occur before or after surgical resection.
5. Patients with Hirschsprung disease need long-term follow-up and care with an interdisciplinary team and may require a bowel management program.

1. Background:

    a. Hirschsprung's disease (HD) is a congenital absence of ganglion cells in the gastrointestinal tract. Rectal involvement is universal, but the length of the affected intestine varies. Treatment requires resection of the abnormal intestine and pull-through of intestine with ganglion cells. Outcomes are typically

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good postoperatively, but these children can struggle with lifelong colonic motility problems and are best considered treated and not cured.

b. Epidemiology: HD occurs in one in 5000 births. It is more common in males with a 5:1 male to female ratio. It is rare in premature neonates. Associated conditions include trisomy 21, MEN 2A, Waardenburg syndrome, colonic atresia, neuroblastoma, and von Recklinghausen's disease. Genetic defects in Hirschsprung disease have been traced to the RET and EDN pathways. Six percent of cases of HD are familial, but up to 70% have a genetic component to the disease.
c. Associations: HD may also be associated with other enteric disorders. Colonic atresia can occur in patients with HD, but is very rare [1]. These cases represent diagnostic and therapeutic challenges. In multiple case reports, the colonic atresia was the primary diagnosis, presenting in the neonatal period. After treatment of the colonic atresia, these infants still had intestinal difficulties and HD was suspected. It is recommended that in children with colonic atresia, HD should be suspected if they continue to have difficulties stooling after treatment [2, 3]. Anorectal malformations (ARM) have also been reported in patients with HD. The diagnosis of HD is also usually delayed in infants diagnosed with anorectal malformations. A change in bowel caliber in patients with ARM may signify another pathology, including HD. Lack of function of stomas in patients who have had a colostomy creation may also indicate aganglionosis [4]. However, there is concern that biopsies done after surgical correction of ARM may show false-positive aganglionosis. If a patient undergoes a pull-through procedure after surgical correction of ARM, it likely causes permanent fecal incontinence [5]. The co-incidence of HD and ARM is around 2% [6].

2. Pathophysiology:

   a. HD is characterized by the absence of ganglion cells in the distal bowel. The segment without ganglion cells is continuous and ends at a transition zone, most commonly above the dentate line. The lack of ganglion cells results in the inability of the affected bowel to undergo receptive relaxation. There are multiple theories regarding the etiology of this lack of ganglion cells: (1) failed migration of ganglion cells along the gastrointestinal tract, (2) hostile micro-environment causing ganglion cell loss, and (3) auto-immune targeting of ganglion cells.
   b. The length of aganglionic colon varies. The most common transition zone is the rectosigmoid accounting for about 75%. HD can affect longer segments including total colon or total intestinal aganglionosis, which is more severe and has a higher mortality rate.
   c. Histology: Rectal biopsy shows absence of ganglion cells in the submucosal and intermuscular nerve plexuses as well as hypertrophic nerve trunks. There are several stains used to diagnose HD, including S-100, acetylcholinesterase, and calretinin. Acetylcholinesterase will stain the hypertrophic nerve fibers, and calretinin will be absent in Hirschsprung disease patients.

3. Clinical presentation:
    a. The most common presentation of HD in the neonate period is failure to pass meconium. Any term infant who does not pass meconium in the first 24 h of life should be suspected of having HD. Other presenting symptoms include abdominal distention, feeding intolerance, vomiting, fever, and sepsis. In some cases, the infant may have signs of intestinal perforation. On rectal exam, there is often a characteristic blast of liquid stool as the finger overcomes and dilates the spasm in the aganglionic segment. This is typical of short segment disease.
    b. Two-thirds of patients with HD are diagnosed in the first 3 months of life. Those with short segment disease can present later. These infants and children have a history of chronic constipation, abdominal distention, recurrent fecal impaction, rectal prolapse, dehydration, malnutrition, failure to thrive, anemia, and hypoproteinemia. Infants who are breast fed may also present late because the breast milk aids in infant stooling. Once weaned, these patients will often present with chronic constipation or even with initial presentation as Hirschsprung associated enterocolitis.

4. Diagnosis:
    a. Differential diagnosis: There are several conditions in which a neonate may not pass meconium within the first 24 h life. These include meconium ileus, meconium plug syndrome, anorectal malformation, and colonic atresia. These can be differentiated from HD with physical exam and contrast enema. A parental history of cystic fibrosis may also assist in the consideration of meconium ileus instead of HD.
    b. Abdominal x-ray: HD must be suspected when an infant does not pass meconium within the first 24 h of life. Often a plain abdominal x-ray will be ordered first, which typically reveals an abnormal bowel gas pattern and distention of the intestines.
    c. Contrast enema: An unprepped water-soluble contrast enema should be ordered if HD is suspected. Ideally, this is done early in evaluation and prior to any instrumentation or digital exam of the anus and rectum. However, barium gives a better image definition. If a patient has HD, contrast enema will often reveal a transition zone: a focus where the diameter of the bowel changes from narrow distally to dilated proximally (see Fig. 1). This is due to the affected distal bowel having abnormal tonic contractions. Following a rectal exam, a transition zone may not be apparent and the study may need to be repeated. Other indications of HD are an abnormal rectosigmoid diameter ratio (greater than one is normal) and persistence of retained contrast 24 h after the study. The contrast enema can also be used to diagnose meconium plug syndrome (which may be associated with HD diagnosis). If there is no passage of stool even after contrast enema, a repeat abdominal x-ray should be ordered for the next day.

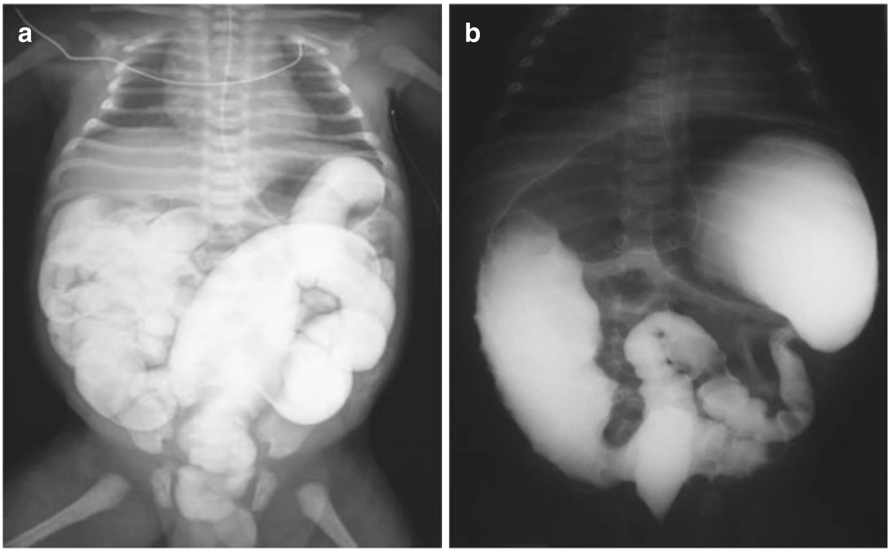

**Fig. 1** (**a**) Subtle transition zone in female infant with Hirschsprung's disease. (**b**) Severe transition zone in male infant with Hirschsprung's disease and Down syndrome. (Source: Michael Allshouse)

   d. Additional evaluation: If there is suspicion of HD and chromosomal syndromes such as trisomy 21, a cardiac echocardiography should be ordered preoperatively [7]. Some institutions recommend ordering a routine cardiac echocardiogram on all patients with HD [8].
   e. Rectal biopsy: The definitive diagnosis of HD is made on rectal biopsy. This will show absence of ganglion cells of the Auerbach (intermuscular) and Meissner (submucosal) plexuses in the wall of the colon. In patients younger than 3 months of age, a bedside suction rectal biopsy is recommended and is effective. The biopsy should be performed 1.5 cm and 3 cm above the dentate line. Rectal suction biopsy can be performed in patients of all ages, but this may need to be done in the operating room for older patients. To ensure adequate depth of specimen, older patients may need a full thickness rectal biopsy. There are very rare complications of suction rectal biopsy including hemorrhage and perforation.
   f. Anorectal manometry: This can be useful in older children to determine if there is a zone of increased pressure and absence of relaxation with proximal distention. Many times, anorectal manometry is the only diagnostic tool for children with ultra-short segment disease.

5. Initial management: Once the diagnosis is suspected, initial treatment includes making the patient NPO and starting intravenous fluids for hydration and resuscitation. Some patients may require a nasogastric decompression if

vomiting or struggling with significant abdominal distention. If signs of Hirschsprung's enterocolitis, perforation, or sepsis are present, broad-spectrum antibiotics should be initiated. Rectal irrigations can aid in clearing the stool and infection.

6. Treatment:

    a. Definitive treatment of HD is resection of the aganglionic segment with pull-through of the normal bowel to the anorectum. This can be performed as a single operation, but some patients may require a temporizing diversion of the bowel if significantly dilated or if the patient is critically ill. Patients with short segment disease who can stool on a regular basis with the use of rectal irrigations can usually undergo a primary pull-through. Patients with long segment disease often require a temporary leveling ostomy. Contraindications to primary pull-through are massive dilation, enterocolitis, perforation, malnutrition, and the inability to determine the transition zone.
    b. A colostomy is performed through a transverse lower laparotomy and the colostomy site is chosen 5 cm above the transition from dilated to narrow colon, typically in the sigmoid. Biopsy with ganglion cells present is the only way to be certain that normal colon has been chosen for the colostomy site. The transition zone may be eccentric, so the colostomy or pull-through should be performed using bowel 3–5 cm above the level at which ganglion cells are identified.
    c. Timing of operation depends on the length of the disease and surgeon preference. Primary pull-through can be performed early or can be delayed several weeks or months as long as a satisfactory stooling pattern can be maintained with dilation or irrigation. Staged pull-through should happen after child has recovered from colostomy and has been able to grow, approximately 3 months.
    d. Types of pull-through: There are several types of pull-through operations (Fig. 2). All operations mobilize the normal bowel with ganglion cells down to the dentate line. The anastomosis is performed 1 cm above the anal canal to retain sensory fibers.

        i. The classic Swenson repair (proctosigmoidectomy) includes resection of the rectum, eversion of the rectal stump outside the anus, and end-to-end anastomosis between the normal colon and distal rectum.
        ii. The Duhamel (retrorectal pull-through) involves anastomosis of the normal colon to the posterior surface of the aganglionic rectal stump. Typically, this is performed with a stapler inserted in the anus to perform a side-to-side anastomosis between normal colon and the aganglionic rectum.
        iii. The Soave-Boley (endorectal) involves a submucosal dissection via the anus to lift the mucosa off the aganglionic rectum. This leaves behind a cuff of muscle above the anus. The cuff of muscle is split posterior to

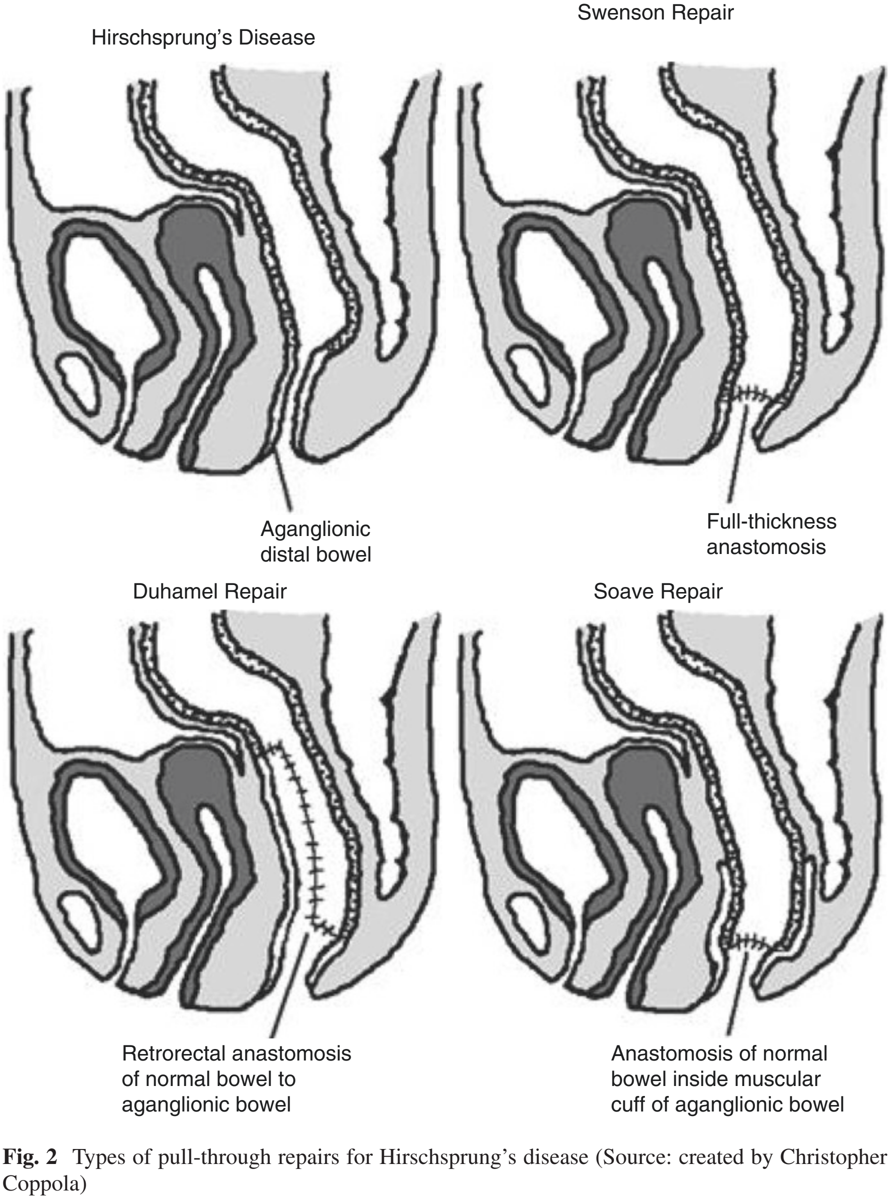
Hirschsprung's Disease
Swenson Repair
Aganglionic distal bowel
Full-thickness anastomosis
Duhamel Repair
Soave Repair
Retrorectal anastomosis of normal bowel to aganglionic bowel
Anastomosis of normal bowel inside muscular cuff of aganglionic bowel

**Fig. 2** Types of pull-through repairs for Hirschsprung's disease (Source: created by Christopher Coppola)

eliminate constriction. The normal colon is brought through this split cuff and anastomosed to the edge of mucosa above the anus.

iv. Pull-throughs can be laparoscopic assisted in which normal colon is identified by laparoscopic muscle biopsy. Abnormal colon is pulled out via the anus and an end-to-end anastomosis is made via transanal approach.

7. Hirschsprung's associated enterocolitis (HAEC):

    a. This is a potentially life-threatening complication of Hirschsprung disease and should always be in the surgeon's mind when treating an infant or child with HD. It is the leading cause of death in children with Hirschsprung disease [9]. It can present either before or after surgery, and it is more common with long segment disease. Infants are thought to be at higher risk of developing HAEC secondary to their immature immune system. In postoperative patients, anal dilations and prophylactic washouts may help prevent HAEC [10].
    b. The pathogenesis of HAEC is still not entirely understood but involves a dysfunctional enteric nervous system causing stasis, overgrowth of certain bacteria secondary to unbalanced intestinal microbiome, and impaired enteric immunity.
    c. Early detection and prevention of HAEC are critical. Prevention includes frequent rectal irrigations to prevent stasis, typically done at home by the parents, as well as use of probiotics.
    d. Typical presentation of HAEC is explosive diarrhea, fever, lethargy, abdominal distention, and in some cases, shock. Plain abdominal radiographs can be used to help aid the diagnosis but should not delay treatment if HAEC is suspected.

        X-ray findings consistent with HAEC include dilated loops of the bowel with a "cutoff" sign (no air distal to the rectosigmoid junction), air fluid levels, pneumatosis intestinalis, and even free air from perforation. Barium enemas should not be used as they could lead to a perforation [11].
    e. Children with suspected HAEC should be given fluid resuscitation and broad-spectrum antibiotics. These patients require close hemodynamic monitoring. Rectal irrigations with warm saline should be started as soon as possible. Some infants with severe HAEC may require urgent diversion with creation of a colostomy. Typical organisms responsible for HAEC are *C. difficile* and rotavirus [12].

8. Hirschsprung disease and Trisomy 21:

    a. Trisomy 21 (Down syndrome) is the most common chromosomal abnormality associated with HD [13]. As a result of this, children with Down syndrome (DS) who struggle with severe constipation should be evaluated for Hirschsprung disease. Coexistence of HD with other congenital anomalies in children with DS must be appreciated. There are documented cases of HD along with congenital anorectal malformation. Newborns with DS and duodenal atresia may have HD that is occult until enteral feeding is established. Any significant feeding intolerance, unexplained abdominal distention, or stooling difficulty should trigger suspicion for HD and the appropriate evaluation performed.
    b. Hirschsprung disease presents similarly in patients with and without Down syndrome [14]. There is some evidence that children with DS may be at

higher risk for preoperative HAEC [15]. Mortality is higher in patients with HD and DS which may be due to common cardiac malformations and immune deficiencies in patients with DS.

c. It was previously thought that patients with DS and HD had worse outcomes. However, this clinical nihilism may not be certain. More recent studies have shown this is not the case, and patients with DS are not at higher risk for a pull-through procedure [16, 17]. They may have delay with toilet training.
d. Definitive surgical treatment of total colonic HD is challenging, especially in children with cognitive and neurodevelopmental delay. Most authorities recommend deferral of colectomy and ileoanal reconstruction until the child can sit on the toilet. This may coincide with toilet training for urinary continence. Children with DS typically take longer to toilet train and a premature colectomy and ileoanal anastomosis in this group can be disastrous. If there is a simple guideline, it is probably to wait and see how these children develop and to consider pull-through only if the child is interested and able to cooperate.

9. Outcomes and long-term management:

    a. The vast majority (95%) of patients with HD survive. Most children have good bowel function but many require laxatives or a bowel regimen. Children may require daily rectal irrigations. Access for antegrade enemas may be appropriate for patients with good response to rectal washouts via an antegrade enema with a Malone procedure or cecostomy tube. Very few children require a permanent colostomy.
    b. Complications after pull-through are constipation, wound infection, fecal incontinence, anastomotic leak, anastomotic stricture, obstruction, enterocolitis, and pelvic abscess.
    c. The majority of pediatric surgeons recommended anal dilations after pull-through. Typically, these begin 2 weeks after pull-through [18]. Pediatric surgeons vary in their recommendations as to how often to dilate, but most agree on once daily. These are done with metal or plastic dilators, and the size is increased weekly by 1 mm.
    d. Chronic constipation is common among patients with HD even after pull-through. These patients often can be managed medically with an aggressive bowel management program including laxatives, or daily enemas. A good bowel management program requires infrastructure in place for timely patient contact regarding symptoms, bowel management questions, or regimen adjustment [19].
    e. Some patients do need reoperation. Typically, these patients develop internal anal sphincter achalasia causing obstruction or incomplete defecation. Repeat rectal biopsy is usually performed as the first step to rule out a retained aganglionic segment. This can be managed with intrasphincteric injection of botulinum toxin A to relax the internal anal sphincter and assist with defecation. For patients with recurrent HAEC or constipation, more aggressive surgical

options may be necessary. Posterior myotomy/myectomy is a viable option, but botulinum toxin injection has become more common [20]. Some patients may require re-do pull-through or ostomy creation. Post-pull-through constipation does seem to improve with age in the general population.

f. Prolapse of the enteric mucosa of the anus may occur after pull-through which may need to be treated by excision of the redundant mucosa or revision of the anastomosis.
g. Bowel management in children with HD often depends on whether the child has colonic hyper- or hypomotility. The universal involvement of the child's rectum and need to resect and reconstruct a "neorectum" is a key bit of information. High amplitude propagating contractions (HAPC) are part of normal colonic motility. They propagate from the right colon to the sigmoid colon, where they terminate. The neorectum lacks the reservoir and sensory capacity of the normal rectum. If HAPCs now propagate to the anus, the child may have significant problems with fecal urgency and even incontinence.
h. Quality of life surveys indicate many patients with HD have lifelong challenges with their diagnosis. This may be related to genetic concerns, not to mention the social, academic, and sexual impact of this congenital colorectal condition. This must be openly discussed with families. Although HD is best thought of as disease that is not cured, the palliation can be exceptional and there are helpful options to help deal with the long-term sequelae.

## Additional Notes

Harald Hirschsprung was a Danish physician who practiced in the late 1800s. He was the first Danish physician specializing in neonatal disease processes. He first described the condition that bears his name as a disease of hypertrophy of the colon. He believed the dilated portion was the diseased portion [21]. These patients would have the dilated bowel resected and an ostomy created. They would do very well and thrive after this. However, once their colostomy was reversed to the nondilated bowel, their symptoms would recur. Orvar Swenson, a pediatric surgeon in Boston, later in the 1940s discovered the true cause of HD—a lack of ganglion cells. He correctly identified the nondilated rectum as the diseased segment and performed the first pull-through procedure, which bears his name, in 1948 [22].

Multiple variations of pull-through procedures have been developed through the years. Boley was able to modify the Soave procedure (which traditionally was a two-operation procedure) to one single operation. With the advent of minimally invasive surgical techniques, laparoscopic pull-through procedures have become more common. There are also transanal pull-through procedures which do not require laparotomy. However, this does present a challenge with long segment disease or total colonic aganglionosis as often mobilization of the necessary length cannot be performed from a transanal approach.

In summary, management of HD has changed drastically as surgical techniques have improved. In the developed world, outcomes for children with HD are quite good. There is still a great need for more resources and surgeons in the developing world for children with HD [23].

## Study Questions

1. An 11-month-old male infant presents to the emergency department with abdominal distention, vomiting, and lethargy. His abdomen is tender, distended, and tympanic to percussion, and he is hypotensive, febrile, and tachycardic. He has a history of Hirschsprung disease and had a Duhamel pull-through within the first 2 weeks of life. Digital rectal exam elicits a large burst of foul-smelling liquid stool. Despite this, patient remains hypotensive and tachycardic. Plain film x-ray shows free air under the diaphragm. What is the most appropriate step in management of this patient?
    a. Fluid resuscitation and IV antibiotics
    b. Contrast enema
    c. Emergently to OR for exploration
    d. A and B
    e. A and C

Answer: (e) This infant is showing signs and symptoms of Hirschsprung's associated enterocolitis. Given his exam findings and vitals, he needs urgent fluid resuscitation and IV antibiotics on the way to the operating room.

2. A 6-month-old female infant presents to the clinic with report of constipation per the mother. Mother states that the patient had one to two stools daily since birth. However, since she has started to wean her from breastfeeding, the infant has been constipated. Mother has reported having to use suppositories to help the infant stool. Infant has no significant medical history and has been growing well. Physical exam reveals a slightly distended abdomen and a normal anus. What is the next step in management of this patient?
    a. Prescribe laxatives
    b. Recommend changing the infant's diet
    c. Abdominal x-ray
    d. Unprepped water-soluble contrast enema
    e. Colonoscopy

Answer: (d) This is a common presentation of Hirschsprung disease that is discovered late. Breast milk makes the stools of infants quite soft and when they are weaned to baby food, the stool consistency changes. In infants with HD, sometimes being breast fed is enough to aid them in stooling for a few months. This infant should undergo contrast enema to rule out HD versus chronic constipation.

3. A term male infant with Down syndrome has repair of a duodenal atresia on day three of life. By day of life 14, the baby is on target feeds of breast milk. After 1 month, he develops abdominal distention and bilious emesis. A contrast enema shows narrow rectum with spasms and a dilated sigmoid colon. You recommend:
    a. Urgent re-exploration for malrotation
    b. Polyethylene glycol laxative added to feeds
    c. Suction rectal biopsy
    d. Upper GI endoscopy
    e. Gastroenterology consult

Answer: (c) This infant has delayed presentation of Hirschsprung disease. This again demonstrates how infants can have associated disorders that delay the diagnosis. Breast milk can also delay diagnosis. Once a contrast enema shows signs of HD, the next step is a rectal suction biopsy for definitive diagnosis. It is highly unlikely that an infant with previous repair of a duodenal atresia would have unrecognized malrotation.

## References

1. Lauwers P, Moens E, Wustenberghs K, Deprettere A, et al. Association of colonic atresia and Hirschsprung's disease in the newborn: a report of a new case and review of the literature. Pediatr Surg Int. 2006;22:277–81.
2. Williams MD, Burrington JD. Hirschsprung's disease complicating colon atresia. J Pediatr Surg. 1993;28:637–9.
3. Kim PCW, Superina RA, Ein S. Colonic atresia combined with Hirschsprung's disease: a diagnostic and therapeutic challenge. J Pediatr Surg. 1995;30:1216–7.
4. Eltayeb AA, Refaiy A. Association of Hirschsprung's disease with anorectal malformations: the early alarming signs for diagnosis and comorbidity related to this association. J Pediatr Surg. 2020;55:1981–3.
5. Vuille-dit-Bille RN, de La Torre L, Hall J, et al. Hirschsprung's disease and anorectal malformations-an uncommon association. J Pediatr Surg. 2021;56:487–9.
6. Nakamura H, Puri P. Concurrent Hirschsprung's disease and anorectal malformation: a systematic review. Pediatr Surg Int. 2020;36 https://doi.org/10.1007/s00383-019-04580-4.
7. Duess JW, Puri P. Syndromic Hirschsprung's disease and associated congenital heart disease: a systematic review. Pediatr Surg Int. 2015;31:781–5.
8. Tuo, G, Prato AP, Derchi M, Mosconi M, et al. Hirschsprung's disease and associated congenital heart defects: A prospective observational study from a single institution. Front Pediatr. 2014;2:99. https://doi.org/10.3389/fped.2014.00099.
9. Pruitt LCC, Skarda DE, Rollins MD, Bucher BT. Hirschsprung-associated enterocolitis in children treated at US children's hospitals. J Pediatr Surg. 2020;55:535–40.
10. Frykman PK, Short SS. Hirschsprung-associated enterocolitis: prevention and therapy. Semin Pediatr Surg. 2012;21:328–35.
11. Demehri FR, Halaweish IF, Coran AG, Teitelbaum DH. Hirschsprung-associated enterocolitis: pathogenesis, treatment, and prevention. Pediatr Surg Int. 2013;9:873–81.
12. Austin KM. The pathogenesis of Hirschsprung's disease-associated enterocolitis. Semin Pediatr Surg. 2012;21:319–27.

13. Stoll C, Dott B, Alembik Y, et al. Associated congenital anomalies among cases with down syndrome. Eur J Med Genet. 2015;58:674–80.
14. Travassos D, van Herwaarden-Lindeboom M, van der Zee DC. Hirschsprung's disease in children with Down syndrome: a comparative study. Eur J Pediatr Surg. 2011;21:220–3.
15. Prato AP, Arnoldi R, Sgro A, Felici E, et al. Hirschsprung disease and Down syndrome: From the reappraisal of risk factors to the impact of surgery. J Pediatr Surg. 2019;21:1838–42.
16. Morabito A, Lall A, Gull S, Mohee A, Bianchi A. The impact of Down's syndrome on the immediate and long-term outcomes of children with Hirschsprung's disease. Ped Surgery Int. 2006;22:179–81.
17. Hackam DJ, Reblock K, Barksdale EM, Redlinger R, et al. The influence of Down's syndrome on the management and outcome of children with Hirschsprung's disease. J Pediatr Surg. 2003;38:946–9.
18. Lin Z, Lin Y, Bai J, Wu D, et al. Outcomes of preoperative anal dilation for Hirschsprung's disease. J Pediatr Surg. 2021;56:483–6.
19. Kilpatrick JA, Zobell S, Leeflang EJ, Cao D, et al. Intermediate and long-term outcomes of a bowel management program for children with severe constipation or fecal incontinence. J Pediatr Surg. 2020;55:545–8.
20. Thakkar HS, Bassett C, Hsu A, Manuele R, Kufeji D, et al. Functional outcomes in Hirschsprung disease: a single institution's 12-year experience. J Pediatr Surg. 2017;52:277–80.
21. Sergi C. Hirschsprung's disease: Historical notes and pathological diagnosis on the occasion of the 100th anniversary of Dr. Harald Hirschsprung's disease. World J Clin Pediatr. 2015;4:120–5.
22. Orvar Swenson MD, et al. New concepts of the etiology, diagnosis, and treatment of congenital megacolon (Hirschsprung's disease). Pediatrics. 1949;4:201–9.
23. Bandre, E, RAF kabore, I Ouedraogo, O Sore, et al. Hirschsprung's disease: management problem in a developing country. Afr J Paediatr Surg. 2010;7:3:166–8. https://doi.org/10.4103/0189-6725.70418.

## *Further Reading*

APSA The pediatric surgery not a textbook (NaT).

# Malrotation

**Alysia A. Agnoni and Christopher P. Coppola**

**Key Points**

1. Malrotation is a congenital anomaly of intestinal positioning that can lead to midgut volvulus.
2. Malrotation with volvulus should be suspected in any child with bilious emesis and requires rapid diagnosis and treatment to avoid the high morbidity of short gut syndrome.
3. Upper gastrointestinal series is the gold standard diagnostic modality, although in some centers sonographic diagnosis is available.
4. Ladd's procedure is the operative treatment.

1. Background
    a. Malrotation is a congenital abnormality of the relative positioning and fixation of organs in the abdomen that occurs between 5 and 10 weeks gestation. It creates the potential for midgut volvulus, a twisting of the intestines which can result in obstruction, ischemia, and catastrophic intestinal loss. Though greater than 50% of volvulus will present within the first month of life, it can present at any age. Volvulus is marked by bilious vomiting and requires immediate detection and surgical correction. The Ladd's procedure was first described by William Ladd in 1936 and remains the standard method of correction. It is described below.

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2. Pathophysiology

   a. Epidemiology: malrotation is present in up to 1 out of 200 births. Unless there are symptoms from volvulus or laparotomy for another reason, it can be clinically silent. Most cases of volvulus occur in the very young, but it has been reported in people as old as their seventies.
   b. Anatomy: with normal rotation and fixation of the midgut, the proximal bowel at the duodenum undergoes a 270-degree clockwise rotation (from the perspective of looking at the child) and the colon undergoes a 270-degree counterclockwise rotation. There is fixation of the duodenum to the retroperitoneum with the ligament of Treitz located in the left upper quadrant of abdomen. There is fixation of the right colon with the cecum in the right lower quadrant and the left colon from the splenic flexure to the left lower quadrant. The mesentery will then arise in an oblique path from the ligament of Treitz to the terminal ileum, in a straight line from the left upper quadrant to the right lower quadrant. This gives the mesentery a broad base which cannot undergo volvulus. In malrotation and non-fixation of the bowel, the mesentery has a narrow base and can easily undergo volvulus around the axis of the superior mesenteric artery. Instead of a ligament of Treitz and fixation of the cecum in the right lower quadrant, abnormal Ladd's bands form, connecting the duodenum to the cecum. When volvulus occurs, it usually happens in a clockwise fashion. With one rotation of the midgut there is obstruction of the lumen, with further twisting, the blood supply is compromised, and ischemia of the midgut occurs (Fig. 1).
   c. Situs inversus: with situs inversus and heterotaxy syndrome (isomerism), there is often malrotation and non-fixation. Other abnormalities may be present such as asplenia and central globular liver. Of note, when situs inversus is present, volvulus will usually occur with a counterclockwise rotation of the midgut, which differs from the volvulus when the organs are on the usual side of the body.
   d. There is much variation in malrotation between individual patients. Different segments of the midgut may be free to undergo volvulus and sometimes there is partially complete fixation of the bowel. As an example, even if the cecum is in the right lower quadrant, it does not guarantee that the proximal gut has undergone normal fixation. Some abnormalities of fixation can cause kinked or tortuous arrangements of the duodenum which can result in obstruction.
   e. Malrotation is called the "ubiquitous anomaly" because it is often associated with other congenital abnormalities. Conditions where the midgut does not return to the abdomen during gestation, such as diaphragmatic hernia, gastroschisis, and omphalocele, can result in malrotation and non-fixation of gut.

3. Clinical presentation

   a. The typical presenting symptom of malrotation is bilious vomiting. It is essential that this history is elicited from the child's parents. They should be asked the color of vomitus, and if it is present on clothing or a towel it can be inspected. Any account of bilious vomiting should raise the possibility of malrotation with volvulus and requires immediate evaluation.

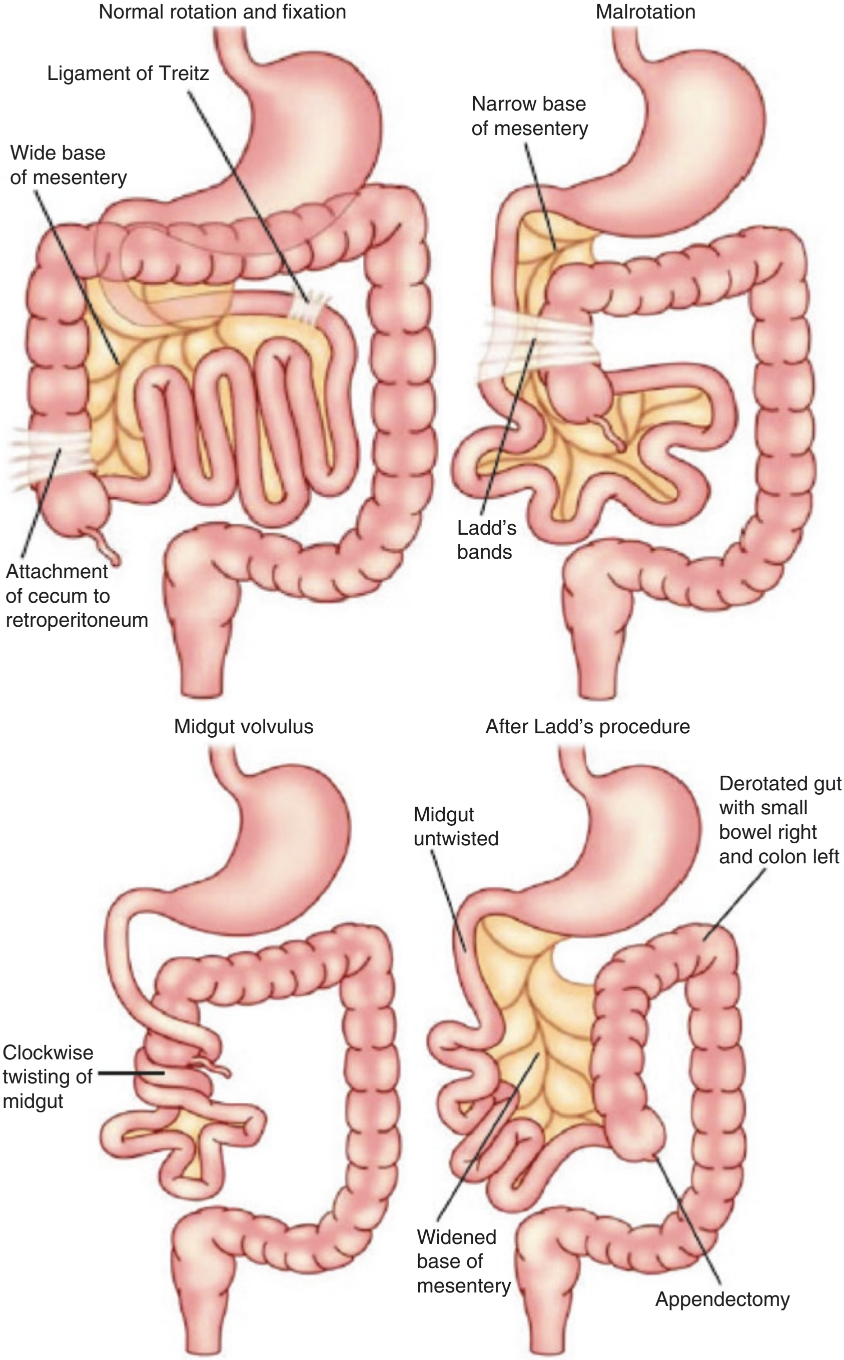
Normal rotation and fixation
Ligament of Treitz
Wide base of mesentery
Attachment of cecum to retroperitoneum
Malrotation
Narrow base of mesentery
Ladd's bands
Midgut volvulus
Clockwise twisting of midgut
After Ladd's procedure
Midgut untwisted
Derotated gut with small bowel right and colon left
Widened base of mesentery
Appendectomy

**Fig. 1** Malrotation and midgut volvulus. In normal development, the midgut is fixed to the retroperitoneum and the mesentery has a wide base. In malrotation, the narrow base of mesentery can undergo volvulus and requires repair with the Ladd's procedure. (Source*:* Christopher P. Coppola)

b. The child may have a scaphoid abdomen resulting from proximal obstruction of the intestine at the duodenum. Inability to pass stomach contents past this point keeps the intestine empty. The abdomen may also present distended if there is perforation and peritonitis. Bloody stool has been described and is often a late sign. If vomiting has gone on for a prolonged period, the patient will be dehydrated, anuric, lethargic, or in hypovolemic shock.

4. Diagnosis: there are many causes for bilious vomiting from infectious to neoplastic, but the cause that must be rapidly excluded is malrotation. Upper gastrointestinal series is the most accurate method to determine if it is present (Fig. 2).

   a. Upper gastrointestinal series: a radiologist experienced in determining rotation of bowel should be consulted. Barium contrast gives the best definition, but water-soluble contrast can be used if there is a chance of perforation to

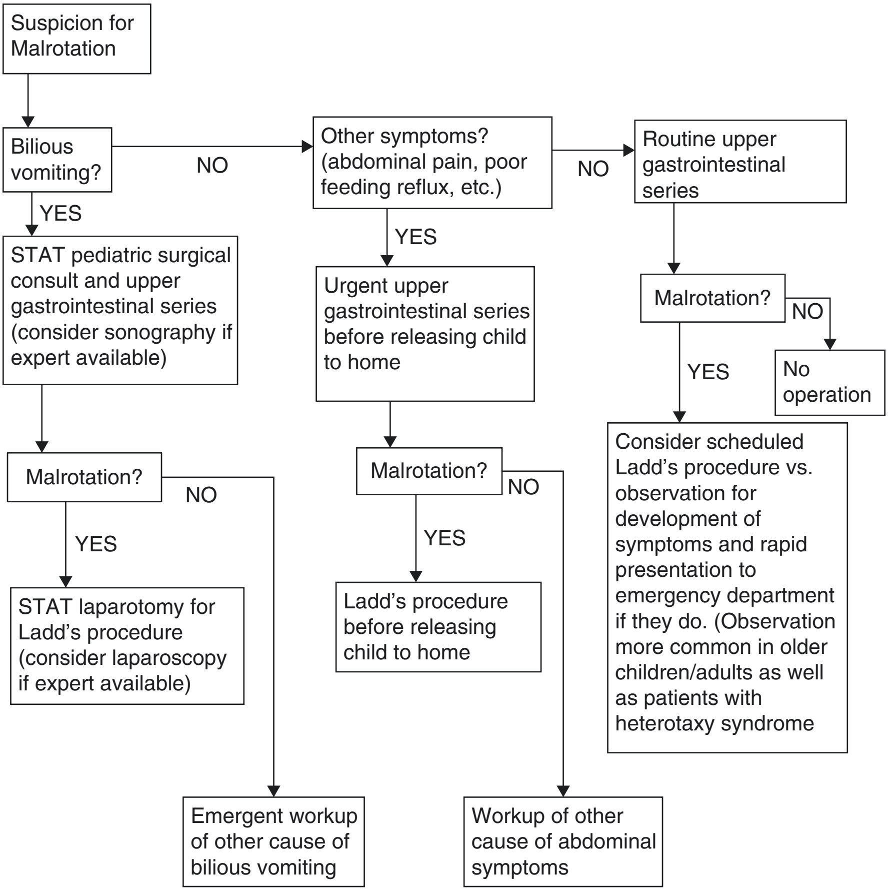
Suspicion for Malrotation
Bilious vomiting?
NO
YES
Other symptoms? (abdominal pain, poor feeding reflux, etc.)
NO
Routine upper gastrointestinal series
YES
STAT pediatric surgical consult and upper gastrointestinal series (consider sonography if expert available)
Urgent upper gastrointestinal series before releasing child to home
Malrotation?
NO
No operation
YES
Malrotation?
NO
YES
Malrotation?
NO
YES
Consider scheduled Ladd's procedure vs. observation for development of symptoms and rapid presentation to emergency department if they do. (Observation more common in older children/adults as well as patients with heterotaxy syndrome
STAT laparotomy for Ladd's procedure (consider laparoscopy if expert available)
Ladd's procedure before releasing child to home
Emergent workup of other cause of bilious vomiting
Workup of other cause of abdominal symptoms

**Fig. 2** Algorithm for the diagnosis and treatment of malrotation. (Source: Alysia A. Agnoni and Christopher P. Coppola)

avoid the irritation caused by extravasated barium. If the child is awake and cooperative, he or she can drink the contrast from a bottle; otherwise, it can be instilled via a nasogastric tube.

    i. There are three criteria for normal rotation of gut. If any of them are absent, the child has malrotation.

        1. Passage of duodenum from left to right, then back again (ligament of Treitz to the left of the spine).
        2. On transverse view, second portion of duodenum is posterior to anterior line of vertebral column.
        3. On anterior view, distal duodenum rises up as high as the level of the greater curvature of stomach or to the level of the duodenal bulb.

  b. Sonography: experienced practitioners can identify the reversed position of the superior mesenteric artery and the superior mesenteric vein. Volvulus can be seen as a whirlpool sign of mesenteric vessels swirled around the superior mesenteric artery. Sometimes interruption of superior mesenteric artery blood flow is seen.
  c. Abdominal X-ray: may show distended stomach and absence of distal gas; however, twisting and untwisting of volvulus can allow some gas to pass distally. If there is a naso-jejunal tube present, the abnormal course of the duodenum can be seen.
  d. Computed tomography: volvulus can be identified as a swirling of mesenteric blood vessels and proximal intestinal obstruction.

5. Initial management: nothing should delay bringing the child to the operating room. When outpatients are discovered to have malrotation, they should be admitted to the hospital. While waiting for operation, a nasogastric tube should be placed and attached to suction. Intravenous fluid is administered, and any electrolyte abnormalities are corrected.
6. Treatment of malrotation and volvulus is the Ladd's procedure.

    a. Components of the Ladd's procedure

        i. Untwist bowel: the bowel usually undergoes volvulus in a clockwise direction (from the perspective of the surgeon). It is untwisted by rotating it in a counterclockwise direction, akin to unscrewing a screw, or turning back time on a clock. In situs inversus, the direction of volvulus is usually reversed.
        ii. Divide Ladd's bands: these are clear adhesive bands binding the duodenum and cecum together. With these organs in close proximity, the mesenteric blood supply is contained in a slender stalk which can easily undergo volvulus. There are variations in the Ladd's bands, and they should be carefully retracted and divided to unwind any kinking of the duodenum and spread the cecum far from the duodenum, which widens the base of the mesentery.

        iii. De-rotation of midgut: small bowel is located on the right side of the abdomen and colon is located on the left. Organopexy is not performed as it is not useful. After operation, adhesions develop which keep the loops of intestine in place.
        iv. Appendectomy: since the appendix will be paradoxically located in the left abdomen after Ladd's procedure, it is removed to eliminate the possibility of future appendicitis with misleading wrong-sided pain. In children younger than 3 months old, an inversion appendectomy can be performed by dividing the mesoappendix with cautery, then inverting the appendix into the cecum with a parotid probe. The last 5 mm of the inverted appendix is ligated, and it auto-amputates into the lumen of the cecum after surgery. This method prevents converting a clean operative field to a contaminated one.

    b. Operative approach: Ladd's procedure can be performed by open or laparoscopic approach. There is some controversy because the laparoscopic approach may not generate sufficient adhesions to keep the bowel in the derotated arrangement after surgery. In the acute setting where volvulus and bowel necrosis are suspected, laparotomy may be the initial method of choice. In cases where volvulus is not clearly diagnosed on UGI, laparoscopy may be attempted initially with the caveat that conversion to open may need to occur if significant bowel resection is warranted.
    c. Short gut syndrome: if the entire midgut is dead when the abdomen is opened, there is controversy in management. Some surgeons, after consultation with the child's family and neonatologist, will offer a course of treatment with comfort as the primary goal. The abdomen is closed without resecting the necrotic material and survival is not expected. However, even with the short gut syndrome that results from removing the entire midgut, there is the possibility of distant future freedom from parental nutrition with bowel transplantation or intestinal lengthening procedures. If there is question about the viability of bowel, a second-look laparotomy in 24–48 h may be a reasonable option.
    d. Asymptomatic patients

        i. There is minimal evidence to support routine screening and/or surgical correction of malrotation in patients with congenital heart disease, specifically those with heterotaxy syndrome [1].
        ii. Consideration for correction should be given to children who are incidentally diagnosed with malrotation. Close observation can be considered in asymptomatic patients, especially in adults.

7. Outcomes

    a. Complications

        i. Loss of major portions of the intestine due to ischemia caused by volvulus results in short gut syndrome and dependence on parenteral nutrition.

ii. The Ladd's procedure intentionally creates intra-abdominal adhesions. Adhesion can result in small-bowel obstruction at any point in the child's future life. Recurrent volvulus can happen if insufficient adhesions do not form from initial surgery.
iii. Death: cases of volvulus which are not treated in a timely fashion will result in midgut ischemia, necrosis, sepsis, and death.

## Additional Notes

The surgical repair of malrotation was included in a publication of surgical diseases in children in 1936, by William E. Ladd, considered to be the father of pediatric surgery. In Ladd's original description, the procedure consists of untwisting intestinal volvulus, dividing the bands between duodenum and cecum to widen the base of the mesentery, and finally replacing the viscera in the abdomen with the small bowel to the right and the large bowel to the left. It was later that additional procedures such as cecopexy or prophylactic removal of the appendix were added. The inversion-ligation method of appendectomy was developed to avoid converting what would otherwise be a clean case to a contaminated case by cutting across the lumen of the appendix as is done in a typical appendectomy.

Although the upper gastrointestinal series has been the gold standard test to diagnose malrotation, there has been interest in using sonography. A recent study showed that sonography for malrotation was diagnostic in 92% of patients tested. For those patients in which sonography determined diagnosis, the positive predictive value and the negative predictive value were both 100% [2]. Sonography can be rapidly deployed for a patient with the urgent symptom of bilious vomiting and does not require ionizing radiation. It does however require the presence of a practitioner with experience with the technique for it to be an effective test.

Most surgeons will use the open technique for performance of the Ladd's procedure to correct malrotation. The laparoscopic Ladd's procedure has been present for years, but several factors have prevented it from becoming the standard of care. Obstruction due to volvulus in some patients will lead to distention, hampering sufficient domain to operate using the laparoscopic technique, and untwisting of midgut volvulus when there has been ischemia can be difficult. There has also been the impression that the laparoscopic method may not produce enough adhesions to adequately prevent future recurrences of volvulus. A recent series of laparoscopic Ladd's procedure showed a low reoperation rate of 6%, which was comparable to a contemporaneous group of children who underwent open procedure, and the conversion to open rate was 14% [3]. The laparoscopic group benefitted from a shorter hospital stay and better cosmesis.

Whenever a completely necrotic midgut is encountered while operating for malrotation and volvulus, the child is facing morbid outcomes and the surgeon has a difficult therapeutic decision. The traditional treatment of closing the abdomen with the necrotic bowel still inside to allow the child to die peacefully is brought into question by advances in treatment for extreme short gut syndrome. In a series of 23 children with short gut after malrotation, 8 achieved freedom from parenteral nutrition through intestinal transplantation and an additional 7 achieved enteral autonomy without transplantation, but for some it took almost 3 years [4]. These results, although hard won, show that there is hope for patients with necrosis of the entire midgut through volvulus. It remains true that a child with midgut volvulus is suffering from a true surgical emergency and those caring for ill children must never forget that bilious vomiting is bad.

## Study Questions

1) Ladd's bands are clear adhesive bands joining ____ and ___ which cause a narrow mesenteric base making it easier for the midgut to undergo volvulus and impede mesenteric blood supply.

   a) The duodenum and cecum
   b) The duodenum and liver
   c) Duodenum and gallbladder
   d) Duodenum and spleen

Answer: (a) The duodenum and cecum. Normal rotation and fixation result in fixation of the cecum in the right lower quadrant, at the lower end of the White Line of Toldt, and fixation of the fourth portion of duodenum/beginning of the jejunum at the ligament of Treitz. In malrotation, these points of fixation fail to form. Instead, the duodenum and cecum are bound together by the Ladd's bands.

2) Which test is the gold standard to diagnosis malrotation?

   a) Abdominal X-ray
   b) Upper gastrointestinal series
   c) Sonography
   d) Computed tomography

Answer: (b) Upper gastrointestinal series. The gold standard test for diagnosing malrotation is an upper gastrointestinal series. It should be performed immediately on a child experiencing bilious vomiting as their presenting symptom so that malrotation with volvulus can be rapidly diagnosed and treated, avoiding bowel loss and short gut syndrome. In some centers, sonography is used as the first-line test to diagnose midgut volvulus, but this requires a practitioner experienced in detecting the sonographic signs of volvulus. An abdominal X-ray can demonstrate that the cecum is in the right lower quadrant, but this does not rule out midgut malrotation and abnormal anatomy of the duodenum. Computed tomography can diagnosis reversed position of the superior mesenteric artery and the superior mesenteric vein, as well as obstruction from volvulus, but it is not the first-line test for malrotation and involves exposure to ionizing radiation without a definitive answer to the question if malrotation is present.

3) Which of the following is not a characteristic of NORMAL rotation on upper gastrointestinal series?
   a) Duodenal contents pass through the C-Loop of duodenum with the ligament of Treitz and the fourth portion of duodenum to the left of the spine before curving down and into the jejunum.
   b) Low-lying pylorus with duodenal contents passing inferior to the stomach in a corkscrew pattern on the right side of the spine.
   c) On transverse view, second portion of duodenum located posterior to the spine in a retroperitoneal position.

Answer: (b) Low-lying pylorus with duodenal contents passing inferior to the stomach in a corkscrew pattern on the right side of the spine. This does not describe a normal rotation as visualized on upper gastrointestinal series and would be consistent with malrotation. For an upper gastrointestinal series to be normal, three characteristics need to be present. (1) There needs to be passage of material in the duodenum from left to right, and then back again to the left, with the ligament of Treitz positioned to the left of the spine. (2) On a transverse view, the second portion of the duodenum is posterior to the anterior line of the vertebral column. (3) On an anterior view, the distal duodenum rises as high as the level of the greater curvature of the stomach or to the level of the duodenal bulb.

## References

1. White SC, Dean PN, McGahren ED, Gangemi JJ, Vergales J. Malrotation is not associated with adverse outcomes after cardiac surgery in patients with heterotaxy syndrome. J Pediatr Surg. 2018;53:1494–8.
2. Wong K, Van Tassel D, Lee J, Buchmann R, Riemann M, Egan C, Youssfi M. Making the diagnosis of midgut volvulus: Limited abdominal ultrasound has changed our clinical practice. J Pediatr Surg. 2020;55:2614–7.
3. Arnaud AP, Suply E, Eaton S, Blackburn SC, Giuliani S, Curry JI, Cross KM, De Coppi P. Laparoscopic Ladd's procedure for malrotation in infants and children is still a controversial approach. J Pediatr Surg. 2019;54:1843–7.
4. Hong CR, Han SM, Staffa SJ, Carey AN, Modi BP, Jaksic T. Long-term outcomes of ultrashort bowel syndrome due to malrotation with midgut volvulus managed at an interdisciplinary pediatric intestinal rehabilitation center. J Pediatr Surg. 2019;54:964–7.

## *Further Reading*

Adams SD, Stanton MP. Malrotation and intestinal atresias. Early Hum Dev. 2014;90:921–5. https://doi.org/10.1016/j.earlhumdev.2014.09.017.

Graziano K, Islam S, Dasgupta R, et al. Asymptomatic malrotation: diagnosis and surgical management an American Pediatric Surgical Association outcomes and evidence based practice committee systematic review. J Pediatr Surg. 2015;50:1783–90. https://doi.org/10.1016/j.pedsurg.2015.06.019.

Millar AJ, Rode H, Cywes S. Malrotation in infancy and childhood. Semin Pediatr Surg. 2003;12(4):229–36.

# Neonatal Abdominal Wall Defects

**Sarah L. Wright and Ryan M. Walk**

**Key Points**

1. Gastroschisis and omphalocele are the most common congenital abdominal wall defects, typically identified on prenatal ultrasound.
2. Fifty to seventy percent of infants with omphalocele have another associated malformation that impacts survival; the survival rate is >90% for those with isolated omphalocele.
3. Semi-analogously, survival for "simple" gastroschisis should be >95%, while "complicated" gastroschisis (those associated with intestinal atresia, perforation, necrosis, or stenosis) will have a less favorable outcome (up to 28% mortality).
4. Operative treatment of gastroschisis varies based on clinical circumstances, institutional resources, and surgeon preference, and can include a variety of options for primary closure or a staged closure using a silo. Bedside closure with a foldover of the umbilical cord stalk to cover the defect can be performed.
5. Omphaloceles are closed primarily when feasible; otherwise, the sac can be allowed to epithelialize via a "paint and wait" approach.

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## Gastroschisis

- Presentation
  a. Evisceration of the abdominal contents through a paraumbilical full-thickness defect in the abdominal wall to the right of the umbilicus.
  b. No overlying membrane, though there may be inflammatory rind from exposure to amniotic fluid.
- Pathophysiology
  a. Failure of closure of the abdominal wall that results in a defect through which the abdominal contents may herniate. Loss of domain from absence of the abdominal viscera within the peritoneum.
- Incidence/epidemiology
  a. Approximately 2–5 per 10,000 live births.
  b. Overall incidence increasing worldwide in recent decades.
  c. Associated with young maternal age, nulliparity, low maternal body mass index.
  d. One-year survival rate is 90–95%.
- Prenatal diagnosis
  a. Most diagnosed on prenatal ultrasound as early as 12 weeks gestation.
    i. Elevated maternal serum alpha-fetoprotein
    ii. Ultrasound findings of intra-abdominal intestinal distention
      1. Greater than ninety-five percent sensitivity and negative predictive value; the absence of fetal bowel distention essentially excludes intestinal atresia.
      2. Specificity approximately 20–40%; the presence of fetal bowel distention is not definitively diagnostic of atresia.
    iii. Close antenatal surveillance recommended as there is a possibility for late gestation fetal demise.
  b. Fetal MRI can be used to aid diagnosis but is not required.
- Initial treatment
  a. Delivery concerns
    i. Some institutions favor delivery at around 36 weeks gestation to both reduce the duration of bowel exposure to amniotic fluid and avoid stillbirth.
      1. Evidence to support this practice is limited.
      2. Similarly, no evidence to support early delivery for the purposes of closing the abdominal defect immediately.
        a. Short duration of silo placement appears to have similar outcomes.

    ii. Nothing precludes vaginal delivery in isolated gastroschisis.
        1. Term vaginal delivery should be the goal for most neonates.
        2. Cesarean delivery reserved for other appropriate fetal or maternal considerations.
  b. Immediate treatment with nasogastric decompression, resuscitation, and broad spectrum antibiotics.
  c. Initial resuscitation should include a 10 ml/kg isotonic fluid bolus followed by administration of maintenance fluids.
    i. Avoid over-resuscitation and consequent bowel edema.
  d. Inspect the bowel for ischemia, perforation, or segments of atresia.
    i. Identification of atretic or stenotic segments can be obscured by the inflammatory peel.
    ii. Conversely, the inflammation can create the appearance of an atresia or stenosis where none exists.
  e. When primary closure is not possible, a silastic spring-loaded silo or staged closure should be used.
  f. TPN will be required for nutrition.
    i. Plan for at least several weeks of durable central access should be coordinated.
        1. Broviac versus Peripherally Inserted Central Catheter (PICC)

- Operative repair
  a. Primary closure
    i. Several options including primary fascial closure, purse-string closure of the skin alone, or reduction of the bowel followed by placement of the umbilical cord remnant over the defect (umbilical turban). This is often performed in the Neonatal Intensive Care Unit. General anesthesia is not required. Intubation for foldover closure is optional based on neonatal consideration and not on the procedure itself.
    ii. Cannot close if replacement of abdominal contents into the cavity results in abdominal compartment pressures in excess of 20 mmHg or peak inspiratory pressures greater than 24 cm $H_2O$.
        1. This could result in decreased cardiac output, respiratory distress, compromise of vascular supply to the bowel, renal failure, decreased perfusion to lower extremities.
        2. Signs of compartment syndrome include decreased urine output, difficulty with ventilation and rising respiratory support, lower extremity edema (decreased venous return), or presence of a firm abdomen.
        3. There should be a low threshold to re-open the abdomen and place a silo if there is any concern for compartment syndrome or bowel ischemia.

    iii. Dilation and irrigation of the rectum to decompress the colon intraoperatively can help with closure by decreasing the volume of bowel to be reduced. Stretching of the abdominal wall between the thumb and index finger can increase intra-abdominal volume.

b. Staged closure with a spring-loaded silastic silo

    i. Infant continues care in the neonatal intensive care unit with IV sedation for spring-loaded silo placement.
    ii. The base of the silo is slipped under the fascial edge and covering the bowel and suspended 90° above the infant. The silo is incrementally tightened as the bowel fills back into the abdomen through a combination of gravity and gentle pressure.
    iii. Care should be taken to ensure adequate perfusion of the bowel throughout the silo reduction process.
    iv. Definitive closure once the silo contents have been completely reduced into the abdomen.
    v. Prior to development of the preformed spring-loaded silo, silastic sheeting was hand-cut and sewn to the fascial edges under general anesthesia. This may be an option if the spring-loaded ring retracts out of the abdomen during serial reductions.

c. "Sutureless closure"

    i. Can be performed either primarily or at the conclusion of a staged silo closure.

        1. If planning on sutureless closure after silo placement, the cord can be placed in the abdomen during the silo reduction to maintain its viability.

    ii. The intestinal contents are reduced into the abdomen and the defect is covered with the umbilical cord and an adherent dressing.

        1. Requires 4–5 cm of umbilical stalk.
        2. Adhesive plastic and non-stick dressings are placed over the cord.
        3. Fascial closure occurs spontaneously as it would with an umbilical hernia.
        4. Epithelialization is typically sufficient at 14 days to prevent leakage of fluid or abdominal contents, at which point the dressing can be removed.

    iii. Offers the potential advantage of repairing the gastroschisis without anesthetizing the neonate.
    iv. Most infants managed with sutureless closure will have an umbilical hernia.

        1. Spontaneously closes as in primary umbilical hernias.
        2. Roughly 10% require umbilical hernia repair.

d. Complicated gastroschisis
    i. Gastroschisis with concurrent intestinal atresia, perforation, ischemia, or stenosis.
    ii. Will typically require longer courses of silo management.
    iii. Five to twenty-five percent of infants with gastroschisis will have associated intestinal atresia.
        1. The vast majority of these will have prenatal ultrasound findings of intestinal distention and polyhydramnios.
        2. The majority of atresias are jejunal or ileal and rarely colonic.
        3. Suspected atresias or stenotic segments should simply be returned to the abdominal cavity and managed expectantly.
            a. Primary repair in the neonatal period may be feasible if the bowel is healthy, without significant rind or edema.
            b. Colonic atresia is a notable exception.
                i. A competent ileocecal valve can create a closed obstruction.
                ii. Such infants ideally receive a colostomy for decompression.
        4. Investigation via contrast studies and re-exploration can be performed at 6–8 weeks if the bowel remains non-functional.
    iv. Prenatal or iatrogenic perforation is managed with creation of a stoma or primary repair.
    v. “Vanishing” or closed gastroschisis is a rare variant where the fascial defect has contracted around the eviscerated bowel, leading to a tenuous mesenteric blood supply.
        1. Management
            a. Careful enlargement of the umbilical fascial defect
                i. Typically, lateral and to the right
                ii. Superior-inferior along the linea alba also acceptable
                    1. Care to avoid injury to the umbilical vessels
            b. Silo placement
                i. Special attention required to avoid volvulus of the narrow vascular pedicle within the silo.
            c. Diligent evaluation of the remaining bowel for ischemia
    vi. Options for large fascial defects
        1. Sequential closure with purse-string sutures
        2. Abdominal component separation
        3. Biosynthetic mesh closure with negative pressure wound therapy
        4. These techniques either help relieve tension so the fascial edges can come together or create an epithelialized surface that can contract over time.

- Undescended testes
  a. Eviscerated gonads are commonly identified in gastroschisis infants.
  b. Ovaries and fallopian tubes are typically returned to the abdomen with minimal difficulty prior to closure or silo placement.
  c. If testicles are identified, orchiopexy is unnecessary in the neonatal period.
    i. Many will spontaneously descend into the scrotum.
    ii. The testicles should be returned to the abdomen adjacent to the deep inguinal rings. This facilitates surgery should orchiopexy prove necessary.
- Post-operative management
  a. Prolonged ileus should be anticipated.
    i. Will require TPN.
  b. Roughly three-week timeframe to advance to full feeds.
  c. Standardized feeding regimens have been shown to hasten the process.
  d. Disorders of intestinal motility may be life-long.
  e. Increased incidence of necrotizing enterocolitis with feeding advancement.

## Omphalocele

- Presentation
  a. Can be immediately classified as one of the following:
    i. Small/medium
    ii. Giant—containing >50% of the liver or a defect of greater than 5 cm.
    iii. Ruptured—10–20% of sacs may rupture prenatally.
- Pathophysiology
  a. Failure of the herniated midgut to return to the abdominal cavity by week 11 of development and failure of midline closure of the abdominal wall at the umbilicus.
  b. The umbilical cord is present on the herniated peritoneal sac.
  c. In the most dramatic presentations, location of the defect can be associated with its etiology.
    i. Cephalic folds: pentalogy of Cantrell—epigastric location.
    ii. Lateral folds: classic omphalocele—central location.
    iii. Caudal folds: cloacal exstrophy—hypogastric location.
  d. Hernia of the umbilical cord
    i. Open umbilical ring, less than 1.5 cm
    ii. Similar entity to omphalocele, more straightforward management

        1. Generally amenable to primary repair.
        2. Can be managed non-operatively in the absence of herniated abdominal contents.

    iii. Associated anomalies may still be present.

        1. Require identical workup to standard gastroschisis.

- Incidence/epidemiology

    a. Approximately 1–4 per 10,000 live births
    b. Associated with multiparity, pre-pregnancy overweight or obese status, advanced maternal age
    c. Higher prevalence in male infants

- Prenatal diagnosis

    a. Most are diagnosed through prenatal ultrasound as early as 12 weeks gestation.

        i. Presence of viscera including the liver outside the abdominal wall after 11 weeks.
        ii. Overlying membrane with significantly shortened umbilical cord inserting directly into the sac.
        iii. Measure size of the defect.
        iv. Once there is a concern for omphalocele, fetal echocardiography and MRI can be considered.
        v. Serial fetal ultrasounds to assess for intrauterine growth restriction.

    b. Fetal echocardiography
    c. Fetal MRI

        i. Not required
        ii. May allow better distinction between liver and bowel in sac
        iii. More accurately determine the extent of liver involvement
        iv. Pulmonary evaluation

            1. Total lung volume: assess for hypoplasia
            2. Ratio of observed to expected lung volume

                a. Less than 50% is indicative of increased postnatal morbidity.

    d. Genetic testing

        i. Associated with an elevation in maternal serum alpha-fetoprotein and beta-hCG.
        ii. Most common chromosomal abnormalities: aneuploidies—trisomy 13 and 18.
        iii. Genetic testing should be completed in all cases where there is even a small concern for omphalocele.

- Evaluation

  a. Up to 80% of infants with omphalocele have another associated malformation.

     i. Thirty-two percent will have congenital heart disease including atrial septal defects, ventricular septal defects, and patent ductus arteriosus

  b. Twenty to thirty percent of infants with omphalocele have chromosomal abnormalities.

  c. Only 20% of cases are isolated.

     i. Greater than ninety percent survival in this group.

  d. Associated abnormalities:

     i. Beckwith–Wiedemann syndrome

        1. Alterations at chromosome 11p15
        2. Macrosomia, hypoglycemia, macroglossia, hemi-hyperplasia, embryonal tumors (Wilms, hepatoblastoma, neuroblastoma, rhabdomyosarcoma), cytomegaly of the fetal adrenal cortex, and renal anomalies
        3. Can exist on a clinical spectrum and have one or many features present depending on specific imprinting anomaly.
        4. Roughly 50% of patients with Beckwith–Wiedemann syndrome will have omphalocele.
        5. Approximately 1 in 10,000–14,000 live births.

     ii. Pentalogy of Cantrell

        1. A constellation of midline defects including abdominal wall, lower sternum, anterior diaphragm, diaphragmatic pericardium, and intracardiac defects.
        2. Cardiac defects range from the most common, ventricular septal defect, followed by atrial septal defect, tetralogy of Fallot, pulmonary stenosis.
        3. Ventral midline defects range from mild sternal cleft to ectopia cordis (an extra-thoracic location of the heart).
        4. Approximately 1 in 65,000–200,000 live births.

     iii. VACTERL

        1. Includes Vertebral anomalies, Anal atresia, Cardiac defects most commonly VSD, Tracheal-Esophageal malformations, Renal aplasia/dysplasia, Radial hypoplasia, and Limb anomalies such as polydactyly/syndactyly.
        2. Omphalocele is an additional associated malformation with the classic VACTERL findings. Other associated abnormalities include hemifacial microsomia, laryngeal stenosis, choanal atresia, single umbilical artery.
        3. Approximately 1 in 10,000–40,000 live births.

    iv. OEIS complex
        1. The associated findings of Omphalocele, cloacal Exstrophy, Imperforate anus, and Spinal defects.
        2. Approximately 1 in 200,000–400,000 live births.
    v. Chromosomal abnormalities
        1. Fetal aneuploidy most common, including trisomies 18 (Edwards syndrome), 13 (Patau syndrome), and 21 (Down syndrome) as well as 45,X (Turner syndrome), 47,XXY (Klinefelter syndrome), and 47,XXX (Triple X syndrome).
        2. Approximately 10% of infants with omphalocele have an associated chromosomal abnormality.

- Initial treatment
  a. Delivery concerns
    i. As in gastroschisis, no specific contraindication to vaginal delivery.
    ii. Special consideration
      1. Giant omphalocele or omphalocele with significant extracorporeal liver may be considered for cesarean section.
      2. Concern for potentially catastrophic liver injury during vaginal delivery.
  b. Care in the neonatal intensive care unit
    i. Intestinal dysmotility is uncommon in omphalocele.
    ii. Enteral feeding can begin earlier than with gastroschisis.
    iii. Additional workup in the perinatal period with serum glucose (to screen for hypoglycemia associated with Beckwith-Wiedemann Syndrome), arterial blood gas, chest radiograph, and echocardiogram (to evaluate for congenital heart disease and pulmonary hypoplasia).
  c. Ruptured omphalocele
    i. Small tears in the membrane can occasionally be repaired with sutures or skin glue.
      1. Undergo eschar formation and epithelization with topical therapy.
    ii. Larger ruptures are more likely to have occurred prenatally.
      1. These may be initially difficult to distinguish from gastroschisis; if there is any liver present in the defect, it is most likely an omphalocele.
      2. Large ruptures require immediate intervention.
        a. A silo can be placed to contain the viscera, similar to gastroschisis.

- Operative repair
  a. Primary repair

        i. Primary closure in omphalocele has similar limitations to those seen in gastroschisis.
            1. Large defects, or those with consequent loss of abdominal domain, will prohibitively raise abdominal compartment pressures, precluding primary closure.
            2. Associated cardiac and pulmonary conditions may contraindicate primary repair.
            3. Particular care must be taken if liver is present within the sac.
                a. Hepatic veins may be present in close approximation to the sac.
                b. Reduction of the liver in the absence of sufficient abdominal domain.
                    i. Compromises hepatic vein or inferior vena cava outflow.
                        1. Liver ischemia
                        2. Cardiovascular collapse
        ii. Closure is achieved with excision of the sac and closure of the fascia over the abdominal contents.
            1. Careful attention to controlling the umbilical vessels within the sac.
            2. Circumferential skin flaps raised over the fascia.
    b. “Paint and Wait” technique.
        i. Use of silver-containing ointments (most commonly silver sulfadiazine) or silver-impregnated dressings and gauze wrapping to promote granulation and epithelialization of the sac. Betadyne is also used.
            1. Silver sulfadiazine may be associated with neutropenia. Need to monitor absolute neutrophil count (ANC).
            2. Immobilization and support of the omphalocele with the gauze dressing can prevent injury.
        ii. Gentle compression applied by dressings aids in sequentially reducing the viscera into the abdominal cavity.
        iii. Eschar formation and epithelialization may take 4–10 weeks.
        iv. Abdominal wall closure is performed when possible without excessive tension and no limitations from associated comorbid conditions.
        v. Several techniques are available when necessary to achieve the needed domain:
            1. Staged closure
            2. Mesh closure
            3. Vacuum-assisted closure
            4. Tissue expanders to develop skin flaps
            5. Component separation of the abdominal wall
            6. Silo closures similar to those seen in gastroschisis have also been described.

vi. The majority of infants with omphalocele will have a ventral hernia that requires repair in the first decade of life.
vii. Wound complications are common after omphalocele repair and include infection, ischemic necrosis of the midline edges, seroma, and recurrent ventral hernia.

| Quick Comparison | Gastroschisis | Omphalocele |
|---|---|---|
| Physical findings | Abdominal defect to the right of the umbilicus with no overlying membrane; umbilical cord in normal anatomical location | Abdominal defect at the umbilicus with overlying peritoneum (unless ruptured) and umbilical cord coming off of sac |
| Embryology | Failure of the abdominal walls to progress or fuse resulting in a defect through which the abdominal viscera can herniate | Failure of rotation of the bowel and return to the abdomen by the 11th week of development |
| Treatment | • Immediate primary closure<br>• Silastic silo<br>• Sutureless closure | • Primary closure<br>• “Paint and wait” technique |
| Associated anomalies | Intestinal atresia | Beckwith–Wiedemann, VACTERL, Pentalogy of Cantrell, OEIS, chromosomal abnormalities |

## Additional Notes

Embryology

Abdominal wall and viscera formation occurs early in embryologic development. At 24–28 days, the foregut, midgut, and hindgut have become increasingly distinct due to cephalocaudal growth of the embryo and the beginnings of lateral closure of the abdominal sidewalls at the foregut and hindgut. The abdominal wall at the midgut remains open and the gut remains in contact with the yolk sac through the broad stalk of the vitelline duct. By the end of the fourth week of development, the stalk is narrowed to the umbilicus and contains the yolk sac duct, umbilical vessels, and the allantois as it enters the embryo and heads caudally to the cloaca. The vitelline duct becomes increasingly narrowed with development, is incorporated into the umbilical cord, and degenerates in the 8- to 12-week timeframe. Persistence of the vitelline duct past this point results in a Meckel’s diverticulum.

Ventral wall defects during development can affect the thorax (ectopia cordis), abdominal contents (gastroschisis), and pelvic organs (bladder or cloacal exstrophy). These are a result of either failure of the lateral sidewalls, or to a lesser extent the cephalocaudal walls, to progress to midline or a defect in fusion once there. With the defect present, the abdominal contents are able to eviscerate into the amniotic cavity.

Physiologic herniation of the bowel through the umbilical ring occurs during the sixth week of development. With rapid growth of the liver and elongation of the primary intestinal loop, particularly from the cephalic limb of the loop about the superior mesenteric artery axis, the abdominal contents outgrow the space in the abdominal cavity and herniate through the abdominal wall through the umbilical ring into the extraembryonic cavity. Once there, the primary intestinal loop rotates 270° counterclockwise (90° during herniation and 180° during return to the abdomen) around the superior mesenteric artery axis, allowing the transverse colon to pass up and over in front of the duodenum. Jejunal and ileal loops make additional small coils, and the cecal bud lies in the right upper quadrant. In the tenth week of development, the liver size is proportionally diminished, the abdominal cavity has enlarged, and the proximal jejunum is the first part of the gut to return to the left hemiabdomen with the remaining loops settling progressively toward the right. The cecal bud is the final part to re-enter the abdominal cavity and with elongation of the large intestine, it subsequently descends to the right iliac fossa. The appendix forms during this timeframe from the narrow cecal bud. Failure of the intestines to return to the abdomen by the 11th week of development results in omphalocele.

## *Complicated Gastroschisis*

In general, longer durations of silo placement are associated with increased disease severity and represent a high-risk subset of neonates. About 5–25% of infants with gastroschisis will have associated intestinal atresia, the vast majority of which will have prenatal ultrasound findings of intra-abdominal intestinal distention and polyhydramnios. These atresias are jejunal or ileal, rarely colonic. Suspected atresias or stenotic segments should simply be returned to the abdominal cavity and managed expectantly, and investigation via contrast studies and re-exploration can be performed at 6–8 weeks if the bowel remains non-functional. Primary repair in the neonatal period at the time of closure would have a high risk of anastomotic complications but may be feasible if the bowel is healthy and without significant rind or edema. In the uncommon variant of colonic atresias with a competent ileocecal valve, a colostomy may be required for decompression. Prenatal perforation or iatrogenic perforation is managed with creation of a stoma or primary repair. “Vanishing” or closed gastroschisis is a rare subset where the fascial defect has contracted around a tenuous mesenteric blood supply to the eviscerated bowel. This is treated with careful extension of the umbilical defect, silo placement with attention to not create a volvulus around the narrow vascular pedicle, and diligent evaluation of the remaining bowel for ischemia. In the event that a fascial defect is too large to be closed, there are strategies to sequentially close with purse-string sutures, use abdominal component separation, or place a biosynthetic material with negative pressure therapy. These techniques either help relieve tension so the fascial edges can come together or create an epithelialized surface that can contract over time.

## Duration of Silo Placement in Gastroschisis

There are equivalent outcomes between patients whose abdomens had been immediately closed primarily and those closed within 5 days after silo placement. The patients who undergo immediate closure or in the first 5 days after short duration silo placement likely represent a favorable presentation of gastroschisis and have similar mortality, time to tolerance of full enteral feeds, duration of TPN, and length of hospital stay. Rate of ventral hernia is about 9% for all repairs but is as high as 12% for both neonates who underwent immediate primary closure or had a long duration of silo placement greater than 10 days. Time to initiation of feeds and time to tolerance of feeds are markers of improved outcomes, and this does not appear to be different for neonates with immediate closure or a short duration silo placement followed by closure. This has significance because it does not necessarily support elective preterm delivery solely to promote immediate primary closure. In general, neonates requiring long durations of silo placement have more complex disease. For this reason, it is difficult to draw firm conclusions from the outcomes of different published gastroschisis cohorts.

## Fetal Intervention in Gastroschisis

There are currently no approved in utero interventions for gastroschisis, but the theoretical goal of fetal intervention in this disease process would reduce damage to the bowel that occurs due to prolonged exposure to irritating amniotic fluid in utero. The same concept has been applied to myelomeningocele with promising success. Damage to bowel in gastroschisis either by the corrosive effect of amniotic fluid or compromised blood supply is one of the leading causes of short gut and need for intestinal transplantation in children.

## Giant Omphalocele

Giant omphalocele is typically defined by large defect size >5 cm or liver evisceration >50% into the sac and generally corresponds to a disproportionate volume between the viscera and the abdominal cavity. These cases not only present challenges for closure but also have associated comorbid physiology including pulmonary hypoplasia or hypertension, systemic hypertension, gonadal anomalies including inguinal hernia and undescended testes, gastro-esophageal reflux, and feeding difficulties. Once diagnosed, giant omphaloceles should be referred to facility that can provide multidisciplinary care. Serial imaging with fetal ultrasound every 4 weeks and fetal MRI at 34 weeks can monitor lung development. Non-stress tests and biophysical profiles can be performed twice weekly starting at 32 weeks. While term vaginal deliveries are possible for most omphaloceles, a cesarean section may be warranted for giant omphalocele. There is no consensus on management of giant omphaloceles, but both staged repair with both "paint and wait" and silo techniques have been described.

### *Associated Anomalies of Intestinal Rotation*

The pathophysiology of abdominal wall defects dictates that essentially all infants with gastroschisis or omphalocele will have malrotation. While a Ladd's procedure can be considered at the time of abdominal wall closure, current consensus indicates that this is not necessary. Nevertheless, families should be counseled regarding the need for immediate evaluation should the child develop bilious emesis at any age.

## Study Questions

Question 1: A neonate is born to a mother who received no prenatal care was found to have a large midline abdominal sac containing loops of bowel. The patient is resuscitated in the neonatal intensive care unit with IVF, started on broad spectrum antibiotics, and the sac is covered with a moist sterile dressing. The patient is hemodynamically stable. What is the next best step in evaluation and treatment?

a. Abdominal ultrasound
b. Echocardiogram
c. Surgical reduction
d. Initiation of TPN

Answer: (b) Echocardiogram. This patient with an unruptured omphalocele has been adequately resuscitated and is stable in the neonatal intensive care unit with appropriate coverage of the defect. As part of the workup and before any surgical interventions are performed, an echocardiogram should be obtained to rule out cardiac defects.

Question 2: Which of the following is not a finding that can be seen with abdominal compartment syndrome caused by excessive tension of wound closure?

a. Decreased urine output
b. Respiratory acidosis
c. Decreased peak inspiratory pressure
d. Lower extremity edema
e. Intestinal ischemia

Answer: (c) Decreased peak inspiratory pressure. Peak inspiratory pressure increases in abdominal compartment syndrome and ventilation becomes difficult as the diaphragm is restricted leading to a respiratory acidosis. Decreased flow to the kidneys and bowel leads to decreased urine output and possible ischemic bowel. Blood flow in the IVC is diminished leading to lower extremity edema.

Question 3: Which of the following is the most common condition associated with gastroschisis?

a. Chromosomal abnormalities
b. Congenital heart disease
c. Premature delivery
d. Intestinal volvulus
e. Intestinal atresia

Answer: (e) Intestinal atresia. Gastroschisis is generally not associated with associated congenital anomalies. Congenital heart disease and chromosomal abnormalities should be suspected in omphalocele but are rare in gastroschisis. Gastroschisis is associated with intestinal non-rotation, but the incidence of volvulus is low. The incidence of intestinal atresia is approximately 10% (see Figs. 1, 2, 3, 4, 5, and 6).

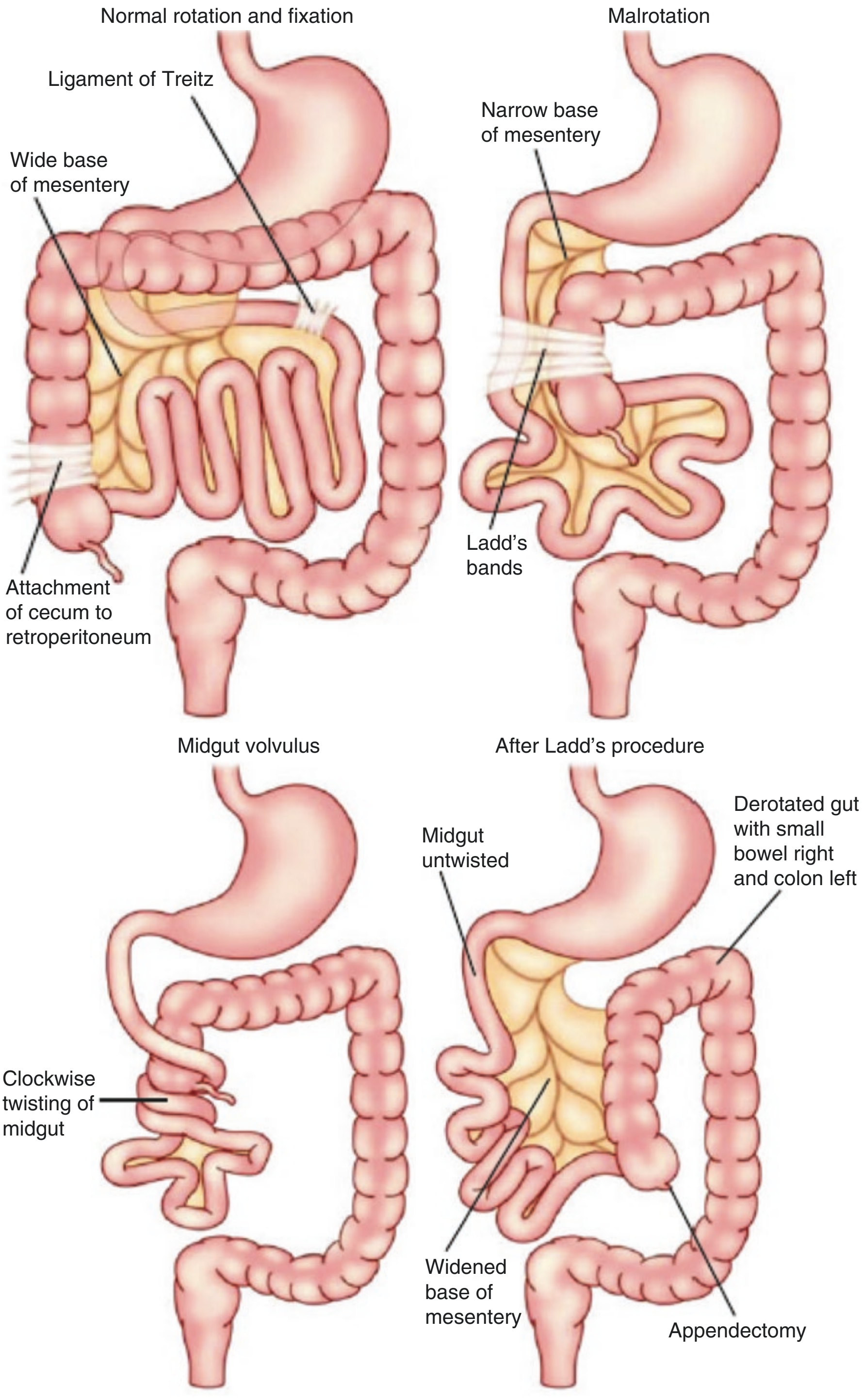

Normal rotation and fixation
Ligament of Treitz
Wide base of mesentery
Attachment of cecum to retroperitoneum
Malrotation
Narrow base of mesentery
Ladd's bands
Midgut volvulus
Clockwise twisting of midgut
After Ladd's procedure
Midgut untwisted
Derotated gut with small bowel right and colon left
Widened base of mesentery
Appendectomy

**Fig. 1** The pathophysiology of abdominal wall defects dictates that essentially all infants with gastroschisis or omphalocele will have malrotation. (Source*:* Christopher Coppola)

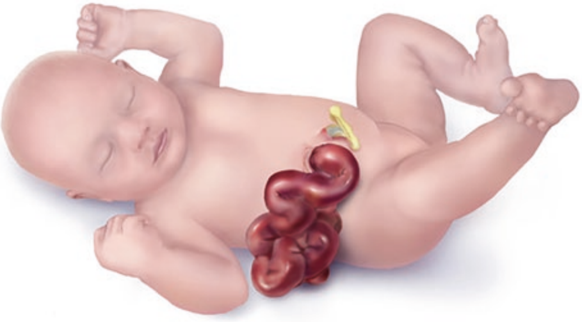


**Fig. 2** In gastroschisis, the defect in the abdominal wall occurs to the right of the umbilicus and the organs outside the abdominal cavity are not covered by a sac. (Source: Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities https://www.cdc.gov/ncbddd/birthdefects/Gastroschisis-graphic.html. Downloaded on 13 July 2021. Image is in the public domain)

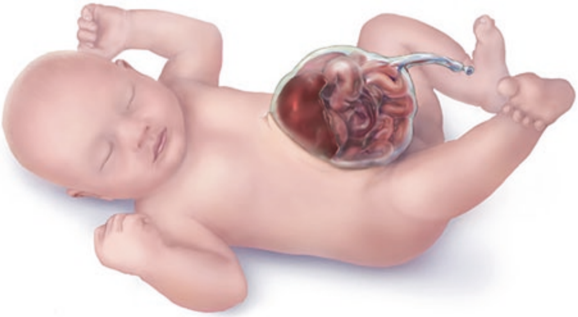


**Fig. 3** Omphalocele with sac covering the organs outside of the abdominal cavity. (Source: Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities. https://www.cdc.gov/ncbddd/birthdefects/images/omphalocele-web_small.jpg. Downloaded on 13 July 2021 Image is in the public domain)

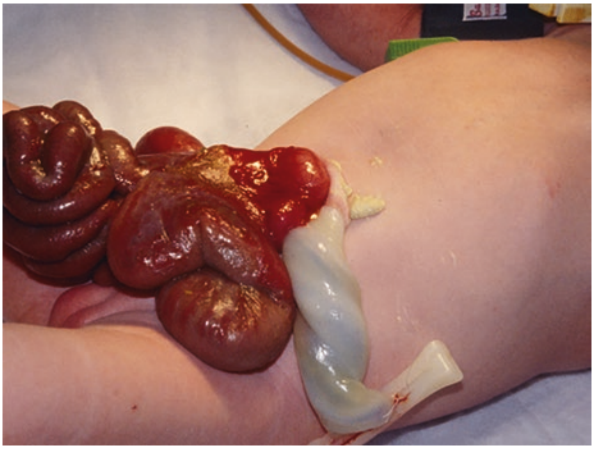
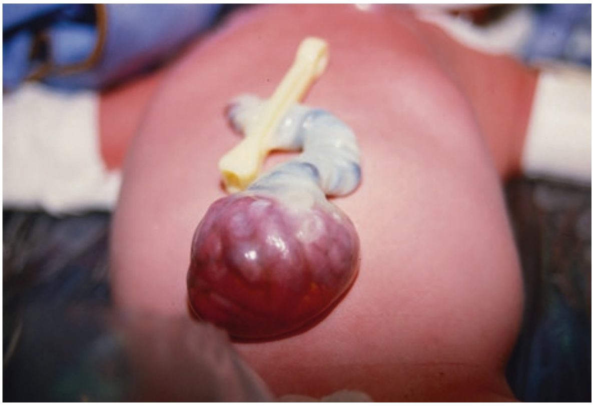


**Fig. 4** Photograph of infant with gastroschisis. (Source: Marc Lessin, MD)

**Fig. 5** Photograph of infant with omphalocele containing small-bowel loops. (Source: Marc Lessin, MD)



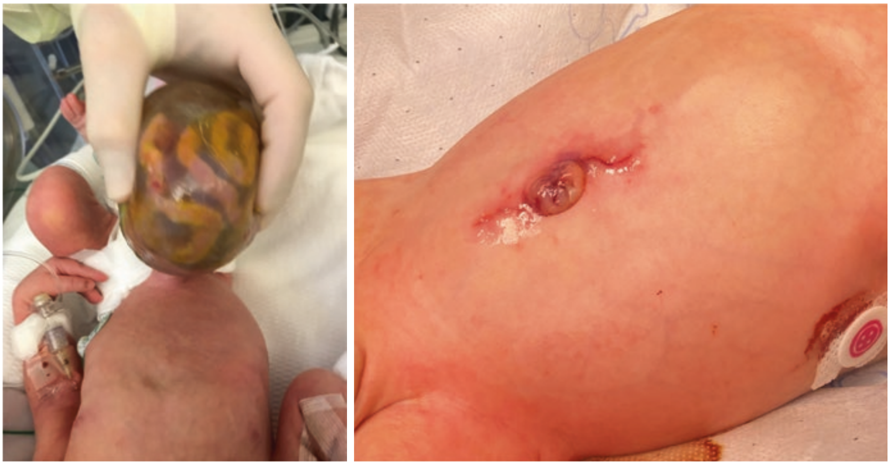


**Fig. 6** Pre- and post-operative photos of a newborn with a hernia of the umbilical cord containing a large volume of intestine. (Source: Ryan Walk, MD)

## Further Reading

Bauman, et al. Management of giant omphaloceles: a systematic review of methods of staged surgical vs nonoperative delayed closure. J Pediatr Surg. 2016;51(10):1725–30.

Benjamin, et al. Anomalies associated with gastroschisis and omphalocele: analysis of 2825 cases from the Texas Birth Defects Registry. J Pediatr Surg. 2014;49(2014):514–9.

Chen. Chromosomal abnormalities associated with omphalocele. Taiwanese Journal of Obstetrics and Gynecology. 2007;46(1):1–8.

Gastroschisis. American Pediatric Surgical Association Prenatal counseling series https://apsapedsurg.org/wp-content/uploads/2020/09/APSA-Gastroschisis-Brochure_FNL.pdf

Ghionzoli, et al. Gastroschisis with intestinal atresia – predictive value of antenatal diagnosis and outcome of postnatal treatment. J Pediatric Surgery. 2012;47(2):322–8.

Hawkins, et al. Immediate versus silo closure for gastroschisis: results of a large multicenter study. Journal of Pediatric Surgery. 2020;55:1280–5.

Omphalocele. American Pediatric Surgical Association Prenatal counseling series https://apsapedsurg.org/wp-content/uploads/2020/09/APSA-Omphalocele-Brochure_FNL.pdf

Stallings, et al. Population-based birth defects data in the United States, 2012–2016: a focus on abdominal wall defects. Birth Defects Res. 2019;111(18):1436–47.

# Anorectal Anomalies

**Laura Tiusaba, Shimon Jacobs, Teresa Russell, and Marc A. Levitt**

**Key Points**

1. Anorectal malformations align across a broad spectrum of complexity, for the surgical challenges involved and the potential to achieve bowel control.
2. Understanding of associated anomalies, particularly urologic and gynecologic, are key, and collaboration with specialists along with devoted nursing expertise involved with the care of these patients, are vital components to achieving good outcomes.
3. The technical reconstruction of the anorectal malformation is clearly an important part of the care of these patients, but long-term treatments, often for many years after successful anatomic repair, are needed to achieve a good functional result.

1. Background
    a. Imperforate anus is a condition affecting males and females in which the anus fails to form in the correct location. There is wide variation in presentation which requires individualization of treatment. Often imperforate anus is accompanied by other anomalies.

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2. Pathophysiology
   1. Epidemiology
      1. Incidence is 1 in 5000 births.
   2. Classification
      1. Male
         1. Anal stenosis: a normally located anal opening within the sphincters but with narrowed diameter (Fig. 1).
         2. Rectal atresia: similar to anal stenosis with a funnel-like anal canal in the center of the sphincters but with an atretic distal rectum (Fig. 2).
         3. Perineal fistula: a connection from the rectum to the skin of the perineum. Sometimes meconium can track subepithelially in the scrotal raphe (Fig. 3). Associated urological anomalies may include penoscrotal transposition, severe hypospadias, and bladder exstrophy.
         4. Normal-sized anal opening in an anterior position: anus forms anterior to the normal location but is still within the anal sphincter muscle complex, accompanied by a relatively small perineal body. This is a normal variant that requires no intervention.
         5. Rectourethral fistulae: In the diagram below the parts of the urethra are represented by the statue's arm (Fig. 4).

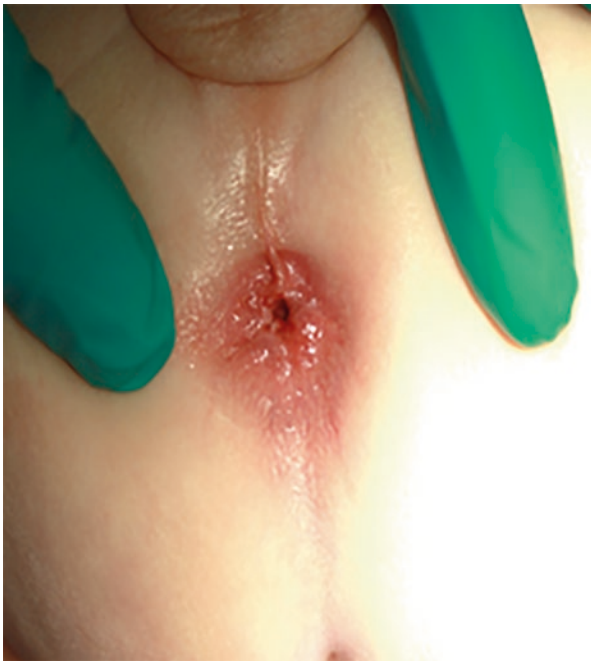


**Fig. 1** Anal stenosis in male, external appearance. (Source: Marc A. Levitt, MD)

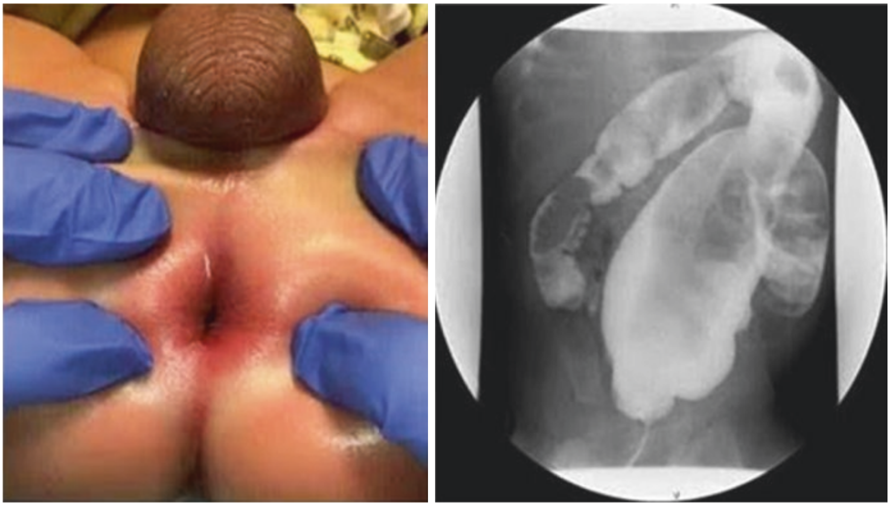
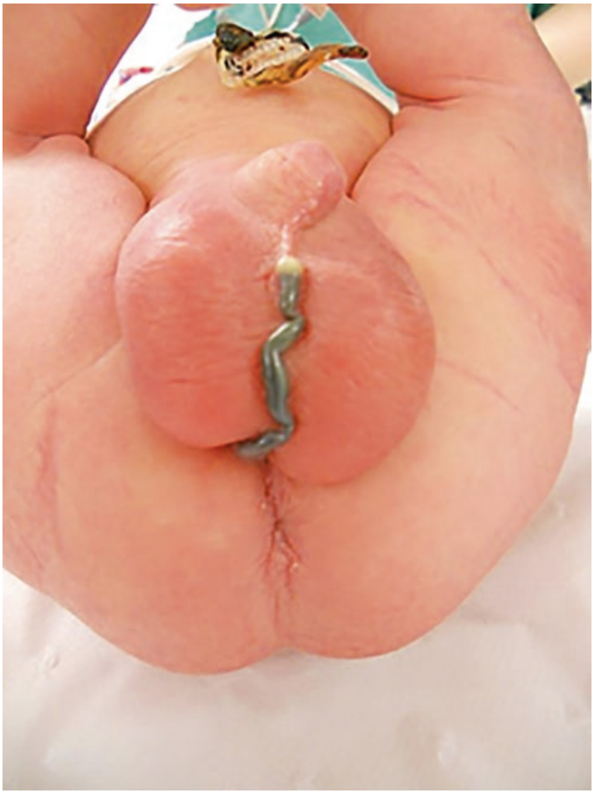


**Fig. 2** Rectal atresia: Left panel shows external appearance. Right panel shows fluoroscopy demonstrating atretic distal rectum. (Source: Marc A. Levitt, MD)

**Fig. 3** Perineal fistula, external appearance, demonstrating subepithelial meconium tract in scrotal raphe. (Source: Marc A. Levitt, MD)



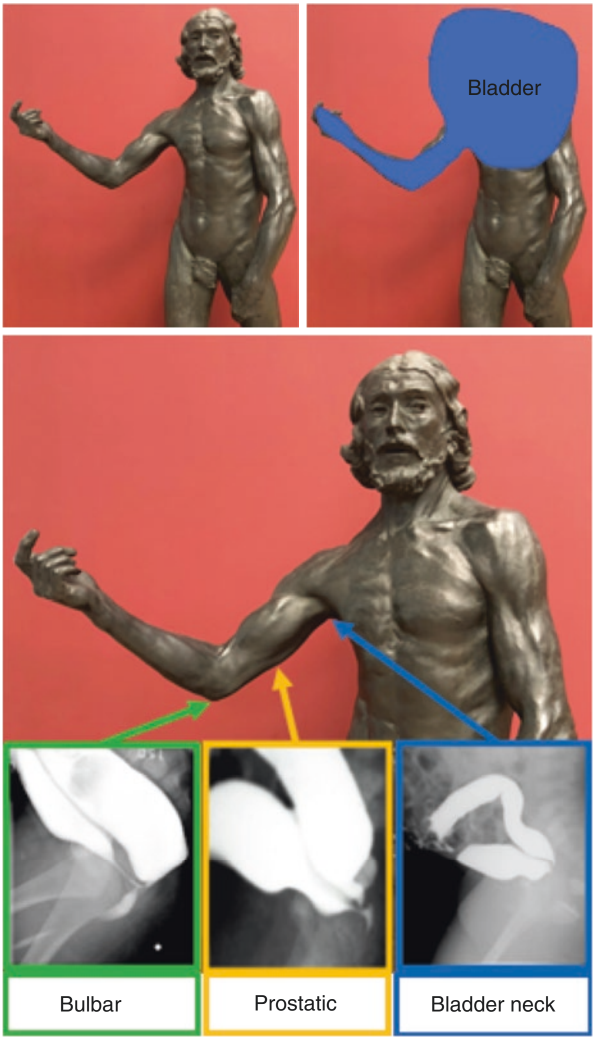


**Fig. 4** Rectourethral fistulae. This photograph of a statue can be used to demonstrate the relation between the fistula and the urethra. The right arm represents the urethra, and the torso represents the bladder seen in a lateral fluoroscopic view. In this schema, a bulbar fistula would join at the elbow, a prostatic fistula would join at the triceps, and a bladder neck fistula would join at the axilla. (Source: Marc A. Levitt, MD)

a. Rectobulbar fistula: a connection between the rectum and the urethra at the level of the bulbar urethra (Fig. 5).
b. Rectoprostatic fistula: a connection between the rectum and the urethra at the level of the prostatic urethra (Fig. 6).
c. Recto-bladder neck fistula: a connection between the rectum and the urethra at the level of the bladder neck. An associated urologic anomaly may include a bifid scrotum. The sphincter mechanism is often close to the base of the scrotum (Fig. 7).

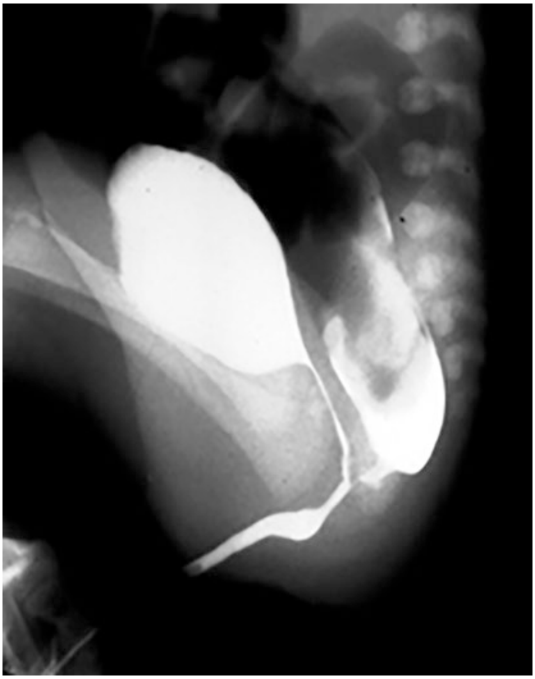


**Fig. 5** Rectobulbar fistula, lateral fluoroscopic view. (Source: Marc A. Levitt, MD)

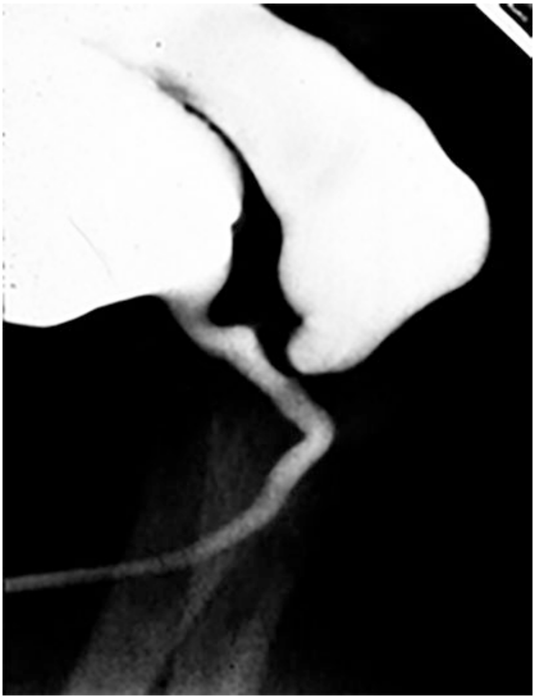


**Fig. 6** Rectoprostatic fistula, lateral fluoroscopic view. (Source: Marc A. Levitt, MD)

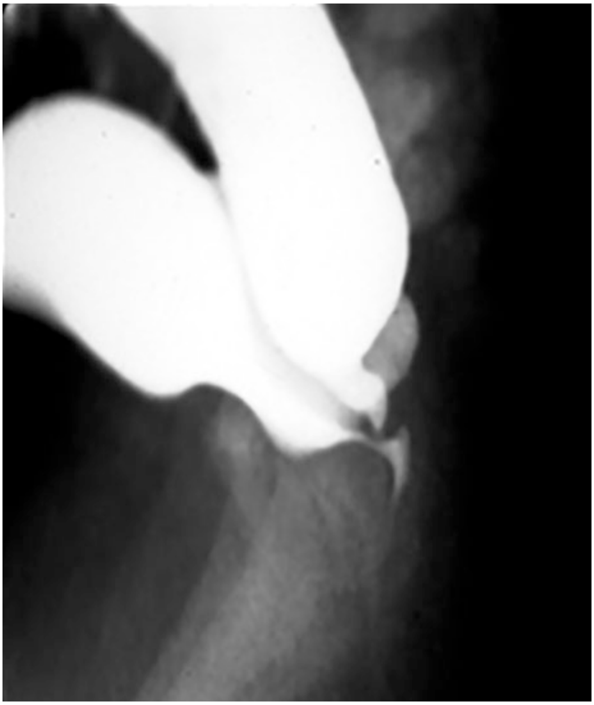


**Fig. 7** Recto-bladder neck fistula, lateral fluoroscopic view. (Source: Marc A. Levitt, MD)

6. Imperforate anus without fistula: congenital absence of the anus and/or perineal opening, without any connection between the rectum and the genitourinary tracts. The distal rectum lies at the level of the bulbar urethra. This is different from a true rectal atresia in which a normal-sized and normally situated anal canal exists but without a connection to the blind-ending rectum. ARM with no fistula defect is commonly associated with Trisomy 21 (Fig. 8).

ii. Female

1. Anal stenosis: normally located anus within the sphincteric ellipse with narrowed diameter (Fig. 9).
2. Rectoperineal fistula: the rectum deviates anterior to the sphincter mechanism in a thin fistula and lies anterior to the sphincteric ellipse (Fig. 10).
3. Rectoperineal fistula is sometimes associated with a mucosal covered perineal body (called a perineal groove) (Fig. 11).
4. An anus in an anterior position: anus forms anterior to the normal location with little or no perineal body, but the anus is still within the anal sphincter muscle complex. This is a normal variant that requires no intervention (Fig. 12).

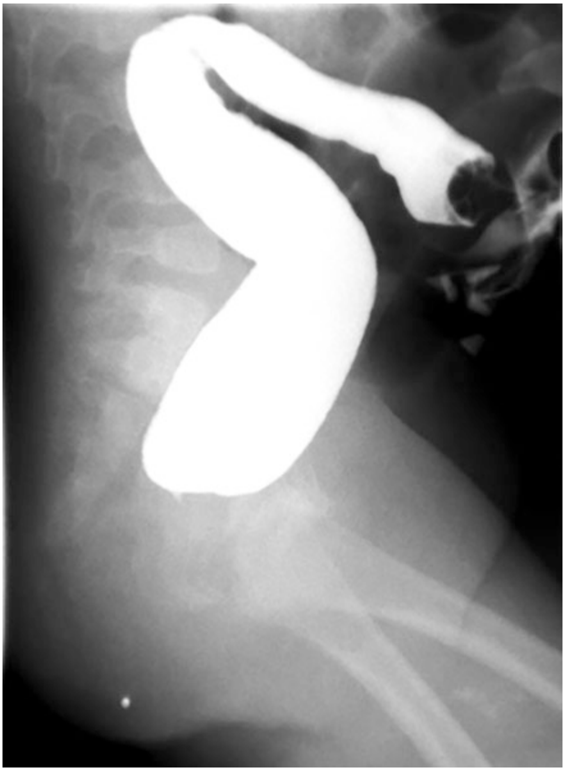


**Fig. 8** Imperforate anus without fistula, lateral fluoroscopic view. (Source: Marc A. Levitt, MD)

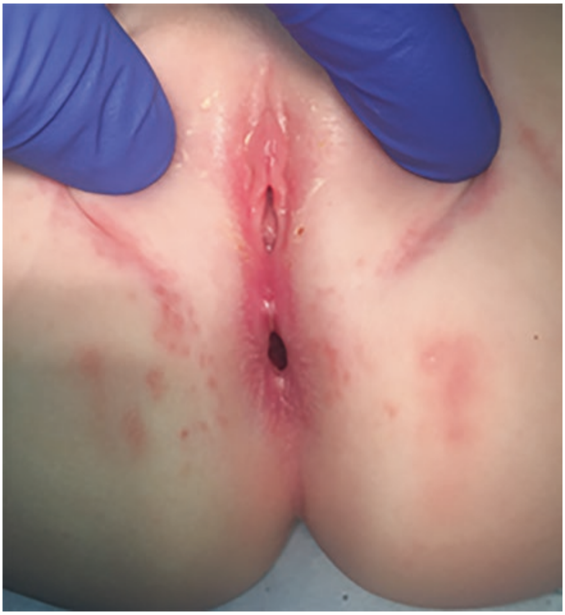


**Fig. 9** Anal stenosis in female, external appearance. (Source: Marc A. Levitt, MD)

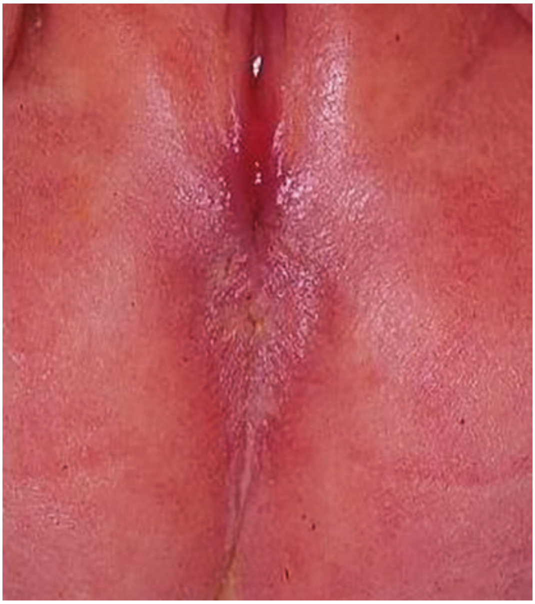


**Fig. 10** Rectoperineal fistula in female. Fistula opening lies anterior to the sphincteric ellipse. (Source: Marc A. Levitt, MD)

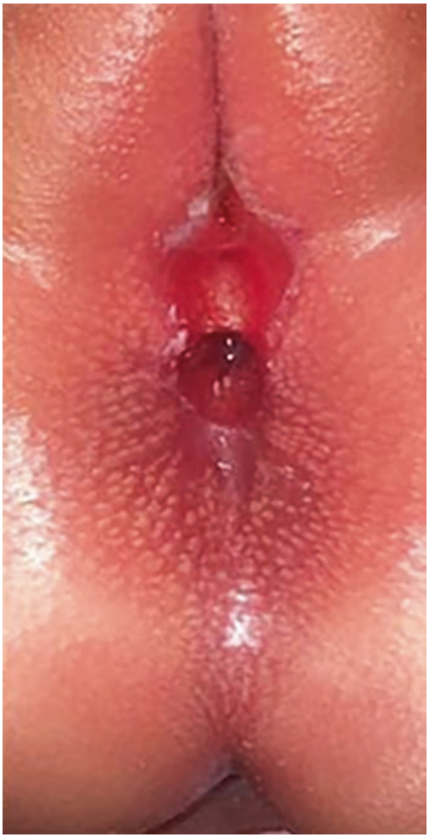


**Fig. 11** Rectoperineal fistula in female, external appearance, demonstrating a mucosal covered perineal body which is called a perineal groove. (Source: Marc A. Levitt, MD)

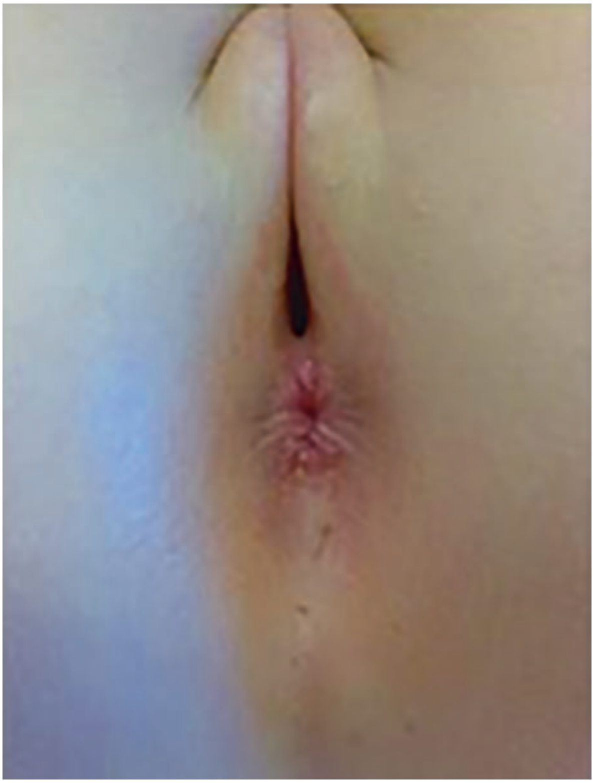


**Fig. 12** Anterior ectopic anus in female, external appearance. (Source: Marc A. Levitt, MD)

5. Rectovestibular fistula: the distal rectum ends in the vestibule, which is within the introitus, but outside of the vagina (Fig. 13).
6. Rectovaginal fistula: the distal rectum ends in the posterior wall of the vagina. It is rare compared to a rectovestibular fistula (Fig. 14).
7. Anorectal atresia without fistula: congenital absence of an anus and/or perineal opening, in a male, without any confluence between the rectum and the genitourinary tracts. ARM with no fistula defect is commonly associated with Trisomy 21.

iii. Persistent cloaca: occurs in females, when the urethra, vagina, and rectum all join to form one common channel. Cloaca is a term which means "sewer" in Latin (Fig. 15).

1. The vagina can become distended and urine-filled from a vaginourethral fistula, also known as hydrocolpos, which can compress the trigone of the bladder, causing ureteral obstruction (Fig. 16).
2. A cloaca can be diagnosed prenatally. In the image below you can see hydrocolpos on a prenatal magnetic resonance imaging (MRI) (Fig. 17).

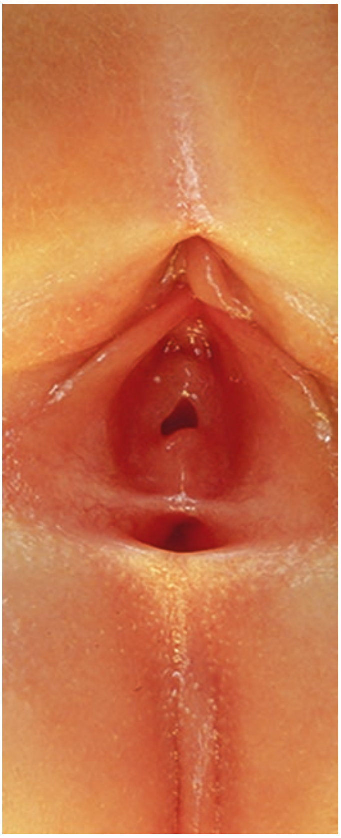


**Fig. 13** Rectovestibular fistula in female, external appearance. (Source: Marc A. Levitt, MD)

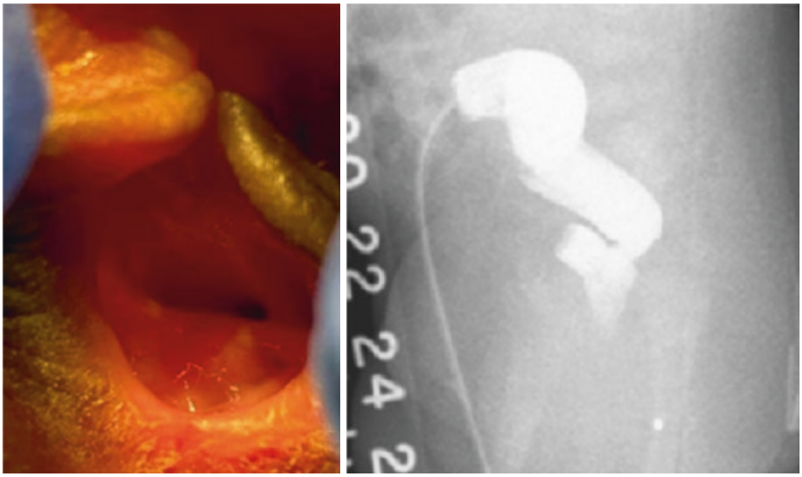


**Fig. 14** Rectovaginal fistula. Left panel: view of vulva showing opening of fistula on posterior wall of vagina. Right panel: fluoroscopy demonstrating rectovaginal fistula. (Source: Marc A. Levitt, MD)

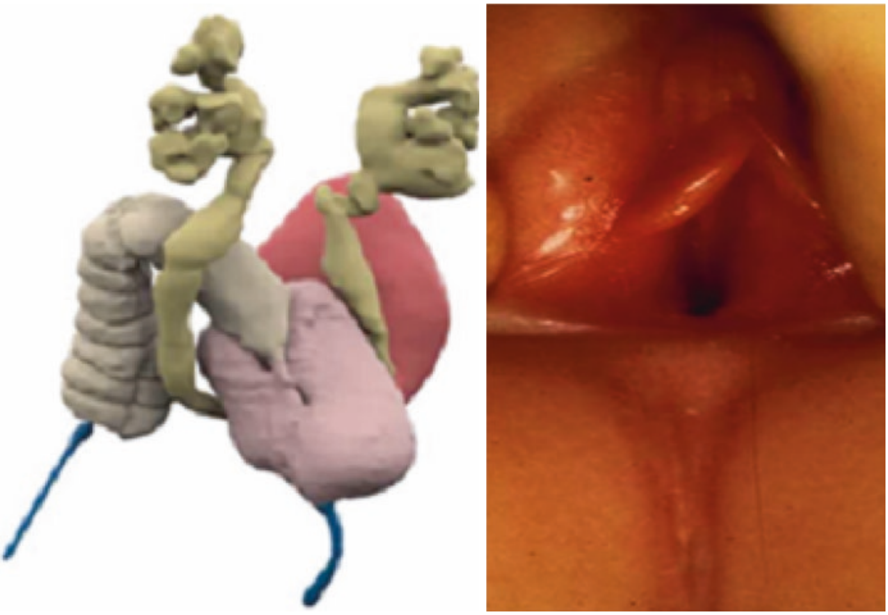
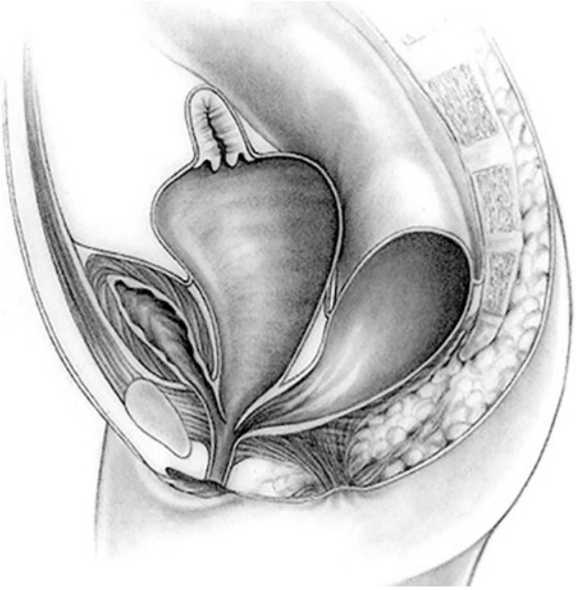


**Fig. 15** Persistent cloaca. Left panel: 3D reconstruction of a cloaca. Right panel: External appearance of persistent cloaca demonstrating single perineal orifice and absence of an anal opening. (Source: Marc A. Levitt, MD)

**Fig. 16** Artist's rendition of a cloaca with hydrocolpos. (Source: Marc A. Levitt, MD)



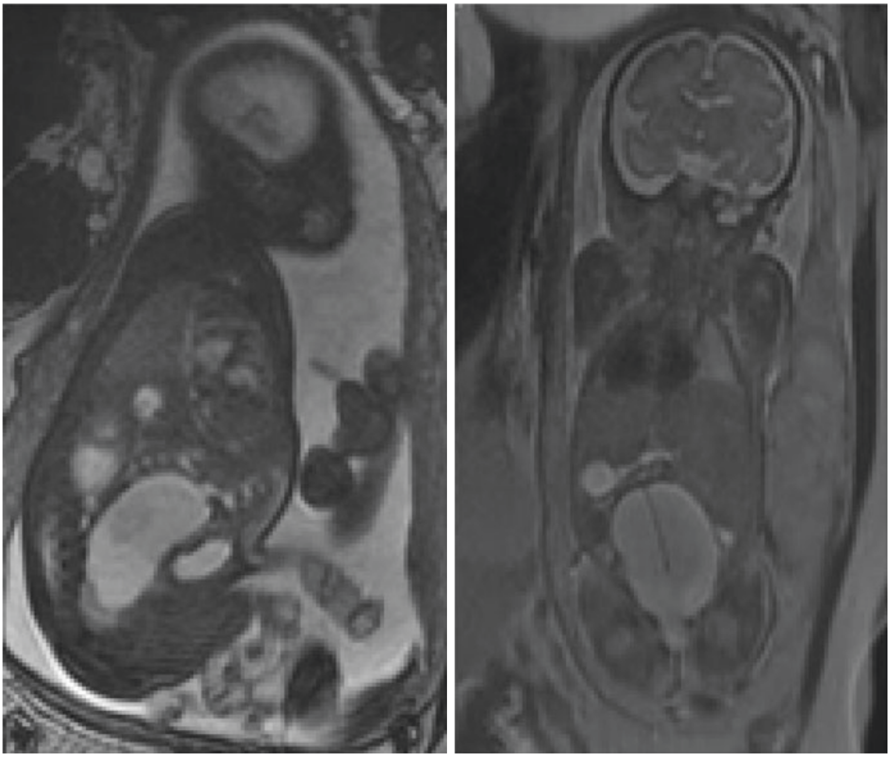


**Fig. 17** Prenatal magnetic resonance imaging (MRI) revealing hydrocolpos with a vaginal septum. (Source: Marc A. Levitt, MD)

            3. Persistent cloaca can be classified by the length of the common channel and the length of the urethra: longer than 3 cm is considered a long common channel and a 1.5-cm urethra is considered a normal length urethra (Fig. 18).
        - iv. Cloacal exstrophy: a complex combination of malformations in males or females in which there is a defect of the abdominal wall with an exposed inner surface of two hemi-bladders separated by an intestinal cecal plate with colonic mucosa and two appendices. There is also a defect in the pubic bone and usually an omphalocele (Fig. 19).
- c. Associated conditions
    - i. VACTERL constellation of anomalies
        1. Vertebral—e.g., scoliosis, hemivertebrae, sacral hypodevelopment
        2. Anorectal
        3. Cardiac—e.g., VSD, ASD, tetralogy of Fallot

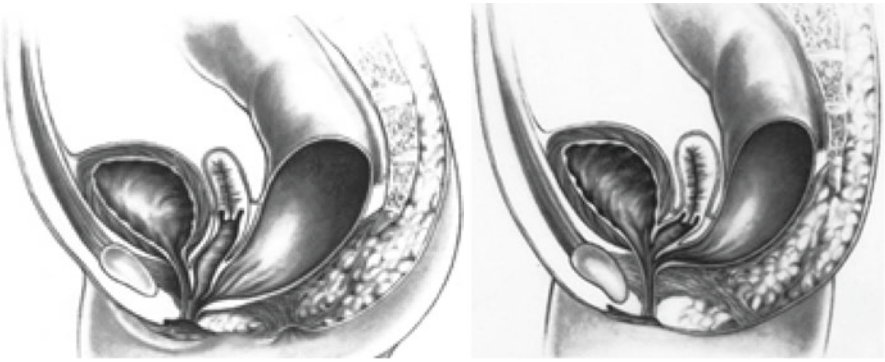
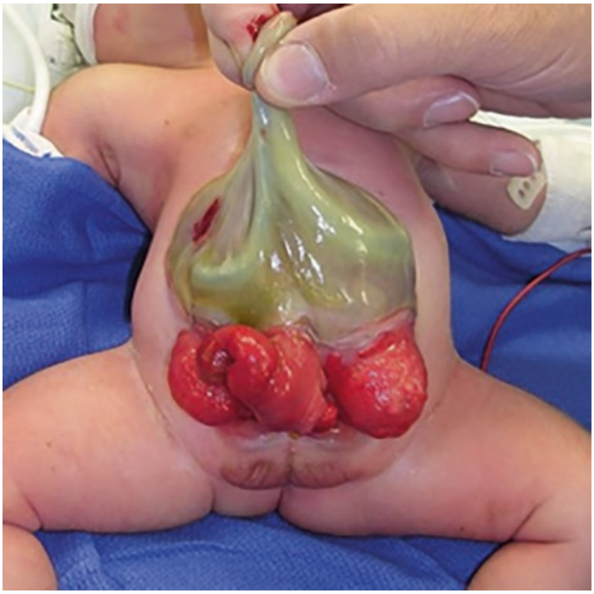


**Fig. 18** Persistent cloaca. Left panel: Short common channel persistent cloaca has a common channel length ≤3 cm and a normal length urethra. Right panel: Long common channel persistent cloaca has a common channel length >3 cm and a short urethra. (Source: Marc A. Levitt, MD)

**Fig. 19** Cloacal exstrophy, external appearance. (Source: Marc A. Levitt, MD)



    4. Tracheo-esophageal fistula
    5. Renal
    6. Limb—absent radius

3. Clinical presentation
    a. History
        i. No passage of meconium or passage of meconium through a fistula.

   ii. In males, urine may contain stool or bubbles passing from the distal rectum, through the fistula, into the urethra or bladder.
   iii. Prenatal diagnosis based on dedicated fetal imaging showing bowel dilation or VACTERL-association anomalies.

b. Physical examination

   i. Anal examination

      1. No anus present or there is an obvious fistula.
      2. Evaluate the sacrum which can be short or absent.

         a. Calculate the sacral ratio using the lateral projection as shown on the images below (Fig. 20). A ratio greater than 0.7 is normal and predicts good sphincters and anal innervation with potential for continence (Fig. 21).

      3. Check for gluteal cleft: a smooth, "flat bottom" indicates a higher lesion.
      4. May have dimple or puckering at the site of the muscle sphincter complex under the skin (connotes good potential for bowel control).

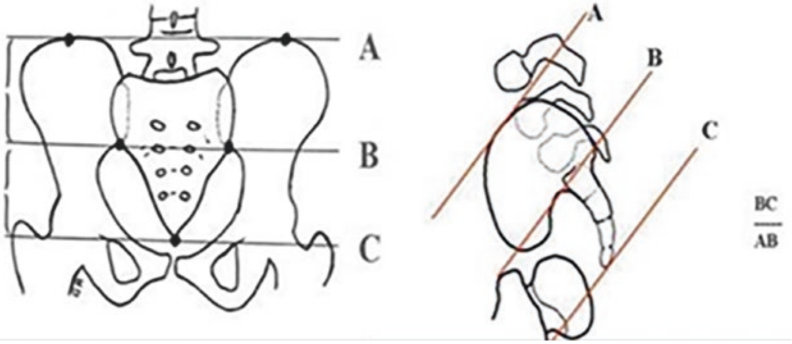


**Fig. 20** Sacral Ratio. A: Top of iliac crest. B: Bottom of sacroiliac joint. C: Tip of coccyx. The Sacral Ratio is calculated by dividing measurement BC by measurement AB. The lateral view is more accurate as the anteroposterior (AP) calculation can be affected by the tilt of the pelvis. (Source: Marc A. Levitt, MD)

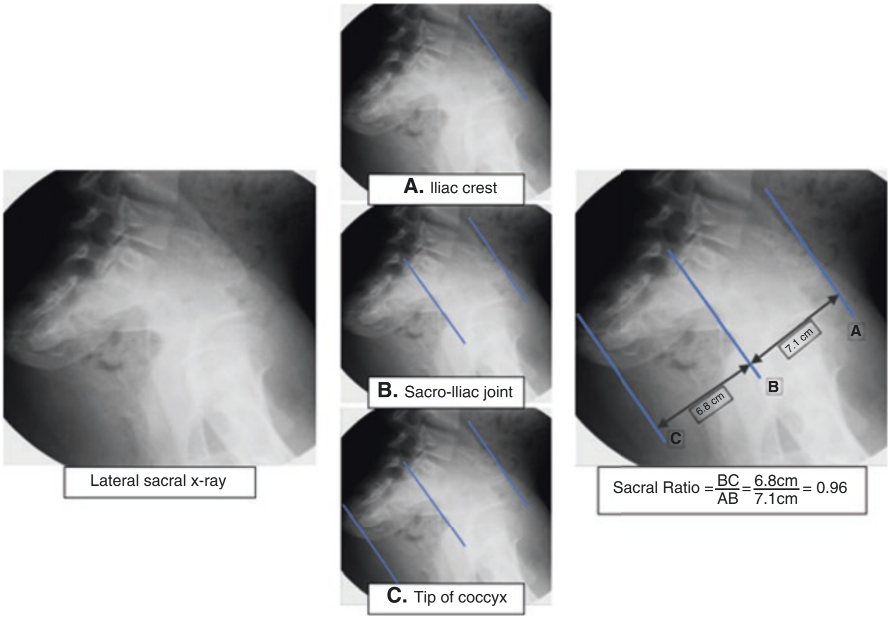


**Fig. 21** Example of normal Sacral Ratio. A: Top of iliac crest. B: Bottom of sacroiliac joint. C: Tip of coccyx. The Sacral Ratio is calculated by dividing measurement BC by measurement AB. In this example, the calculated Sacral Ratio of 0.96 is greater than 0.7 which predicts good future sphincter function and potential for continence. (Source: Marc A. Levitt, MD)

        5. Low lesion, like perineal fistula, may have meconium or mucous “pearls” visible through the thin subepithelial layer of skin (Fig. 22).
        6. “Bucket-handle” flap of skin may be present, commonly a rectoperineal fistula opening will be just under the skin in this case (Fig. 23).

    ii. Examine the abdomen for masses, distended hollow structures (colon, bladder, vagina), and abdominal wall defects
    iii. Evaluate for associated anomalies

        1. Check limbs for absent radius, clubbed foot, or other anomalies
        2. Check spine and sacrum
        3. Listen for cardiac murmurs
        4. Check for physical findings associated with Trisomy 21 such as upward slanting eyes, low-set ears, Simian crease of the palms, etc.

4. Radiologic evaluation

    a. Chest X-ray: check heart and that there is no esophageal abnormality preventing the nasogastric tube from passing into the stomach.
    b. Spinal X-ray: check for abnormal or missing vertebrae.
    c. Echocardiography: check for structural heart abnormalities, presence of pulmonary hypertension, or significant shunting.

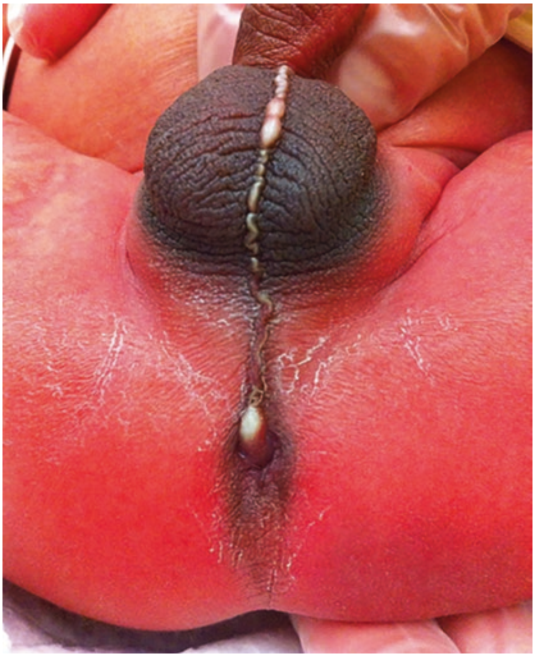


**Fig. 22** Perineal fistula, external appearance, exhibiting mucus "pearls" below the epithelium of perineal skin. (Source: Marc A. Levitt, MD)

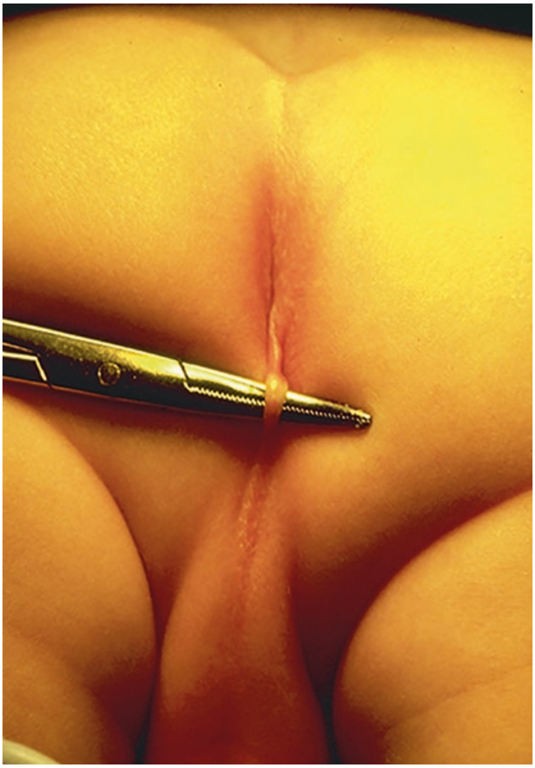


**Fig. 23** Perineal fistula, external appearance, exhibiting a "bucket handle" of skin at the site of fistula. (Source: Marc A. Levitt, MD)

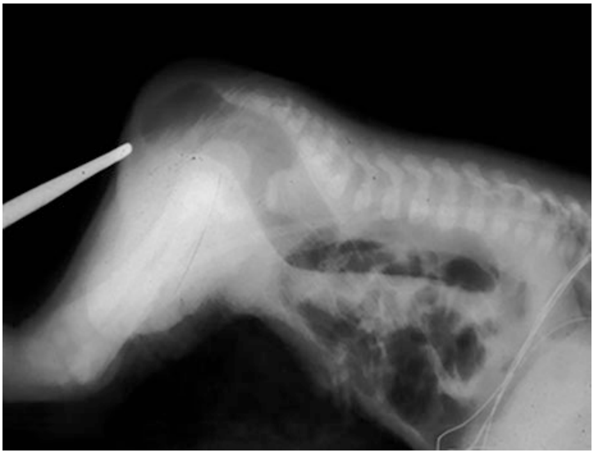


**Fig. 24** Cross-fire film. This radiograph is a transverse lateral view of a baby in prone position with buttocks raised. The blind-ending rectal stump fills with air and is demonstrated. The opaque marker identifies the expected external location of the anus. (Source: Marc A. Levitt, MD)

   d. Renal and abdominal sonography: check for renal abnormalities, distention of bladder and collecting system or pelvic organs, or masses.
   e. Spinal ultrasound: check for tethered cord.
   f. Abdominal sonography: to identify obstructed or distended abdominal structures, hydronephrosis, absent kidney, hydrocolpos (in the setting of cloaca).
   g. If a primary repair is considered, a transverse lateral film with the child in prone position with hips flexed to raise the buttocks can sometimes show air in the blind-ending rectal stump and give information about the height of the defect and distance from the expected location of the anus, marked externally with a radiopaque object (Fig. 24).
   h. Vaginoscopy: to identify any associated Mullerian anomalies (vaginal septum, distal vaginal atresia, etc.)
   i. Cystoscopy: to identify any associated urological anomalies, measure urethral length, and common channel in cloaca patients, and to obtain a urine sample for testing
   j. 3D Cloacagram: catheterization and contrast injection of the bladder, vagina, and distal rectum (via mucous fistula after colostomy creation) and CT images are obtained to reconstruct three-dimensional images essential for surgical planning of a cloacal malformation reconstruction (Fig. 15).

5. Treatment in the newborn period

   a. IV fluid, nothing by mouth, and nasogastric tube decompression
   b. If a fistula is visible in the perineum or vagina, it can be dilated and irrigated to relieve obstruction of the colon by thick meconium. Primary repair in the newborn period or delayed by 1–3 months is possible in such a case.
   c. May need to time treatment around care for other anomalies, such as trachea-esophageal fistula or cardiac defects.
   d. Anal stenosis can sometimes be treated with serial dilations, but an anal repair is often required. A presacral mass (usually a teratoma) must be ruled out.

e. A perineal fistula can be corrected by surgically relocating the anus more posteriorly to lie fully within the anal sphincter muscle complex. This is performed by a midline incision in the perineum and reconstruction of the anus with the two halves of the sphincter complex surrounding it.
f. Rectourethral fistulas require a diverting colostomy at birth, to divert the stool and allow the baby to feed and grow, and it also provides access for a contrast study through the mucous fistula to identify the height of the rectum and its relationship to the urinary tract.
    i. Open the colostomy in the proximal sigmoid colon: need to leave enough length of colon distal to the stoma so that sufficient length can be mobilized from below to reach the anus for reconstruction.
    ii. A divided stoma or a loop colostomy (Turnbull) will suffice, a consideration is to use an area with fixation to the left retroperitoneum to avoid prolapse, especially if performing a loop. A divided stoma should have the colostomy and the mucous fistula sufficiently distanced to allow for isolated pouching of the colostomy.
    iii. Colostomy can be taken down in a third operation after the anus is reconstructed, and no anal stricture has developed.
g. Posterior sagittal anorectoplasty (PSARP):
    i. Child in prone position, on padding, with hips flexed.
    ii. Locate sphincter muscle complex using electrical stimulator.
    iii. Incise skin in midline and divide sphincter complex in half.
    iv. Locate rectum within ischiorectal fat and levator complex.
    v. Open the rectum and mobilize posteriorly and laterally, then anteriorly until fistula is encountered or in case there is no fistula, until the rectum is separated from the urethra anteriorly. Urinary structures are at high risk of injury while dissecting the rectum.
    vi. Divide and ligate fistula with absorbable suture.
        1. In males, care must be taken not to narrow the urinary tract. A Foley catheter is used to locate and avoid injury to the urethra and bladder.
        2. In females, after closing the fistula to the back of the vulva or vagina, the perineal body is reconstructed as a barrier between the vagina and the rectum.
    vii. The sphincter muscle complex is reconstructed around the rectum as a newly created anus.
    viii. For bladder neck or high prostatic fistulas, a laparoscopic approach for the distal rectal dissection and fistula ligation is employed.
h. Persistent cloaca
    i. All patients need diverting colostomy at birth and drainage of hydrocolpos if there is hydronephrosis. Drainage is usually possible with catheter-

ization of the common channel. See below photographs of bilateral hydrocolpos (Fig. 25) and a case of massive abdominal dilation due to hydrocolpos (Fig. 26), both requiring drainage.

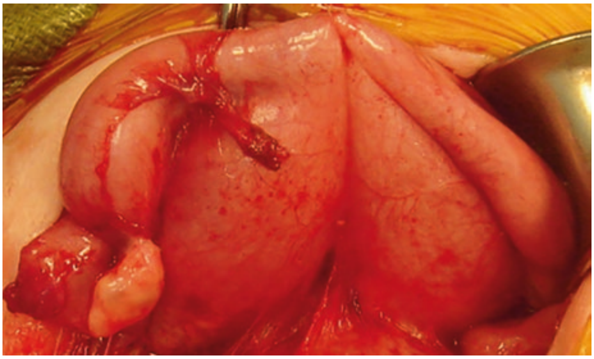


**Fig. 25** Bilateral hydrocolpos in a child with persistent cloaca, which required drainage of the hydrocolpos. (Source: Marc A. Levitt, MD)

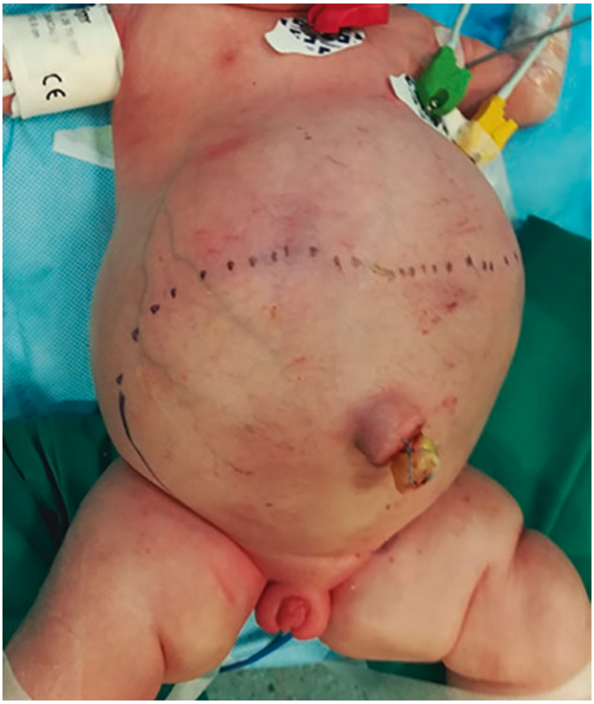


**Fig. 26** Massive abdominal distention due to hydrocolpos in a child with persistent cloaca, which required drainage of the hydrocolpos. (Source: Marc A. Levitt, MD)

    - ii. Less than 3 cm common channel with a urethra greater than 1.5 cm can often be repaired in a single operation using a total urogenital mobilization (TUM) by reconstructing the anal, vaginal, and urethral openings (see figure of cloaca with common channel <3 cm in 2.b.iii).
    - iii. Greater than 3 cm common channel or those with a short (less than 1.5 cm) urethra will require a urogenital separation. In most cases the native vagina reaches, but rarely, a colonic conduit will be required to recreate the vagina. The colostomy should be placed proximal enough so that there is enough colon to construct the anus and to augment the vagina if needed (see figure of cloaca with common channel >3 cm in 2.b.iii).

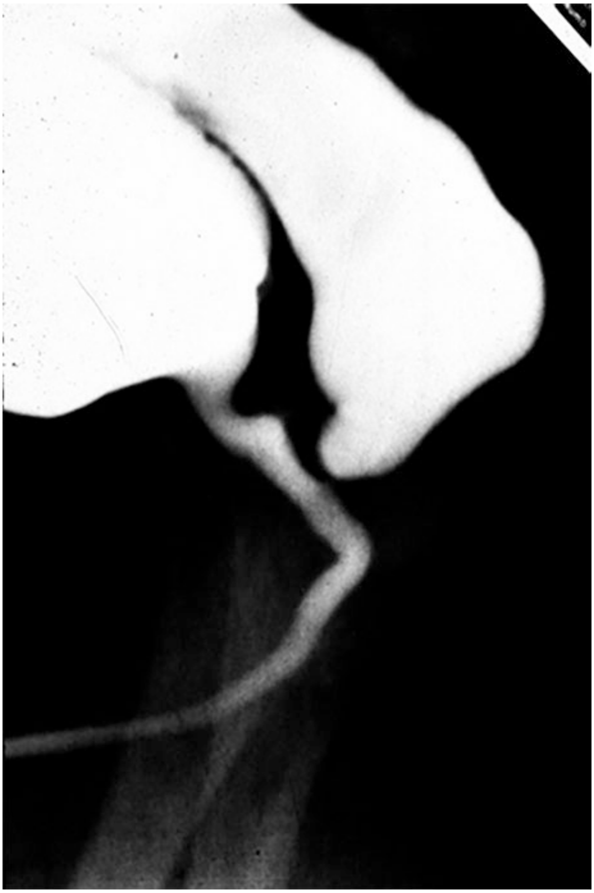
6. Outcome
    - a. Difficulties with stooling are common.
        - i. Patients with high anorectal malformations, a poor sacrum (as classified by the sacral ratio), and associated spinal cord anomalies are likely to have incontinence. They can however be rendered socially continent and clean using a mechanical program of enemas, retrograde or antegrade.
        - ii. Constipation is common and must be aggressively and proactively treated with stimulant laxatives.
    - b. Children may toilet train later than children who have not had a surgically created anus. The vast majority of patients can achieve voluntary bowel movements.
    - c. Regular stooling is attainable for most children, but the majority will require stool regimen, such as daily stimulant laxative or enemas if those are needed to achieve social continence.
    - d. A Malone antegrade colonic enema (MACE) procedure, also known as an appendicostomy, for antegrade flushes is often a helpful adjunct.

## Study Questions

1. What is the clinical value of the sacral ratio?
   It can be used as a predictor of:
    - a. Continence as the bony development correlates with development of perineal nerves and sphincter muscles.
    - b. Location of the rectum during the posterior sagittal approach.
    - c. Associated renal anomalies.
    - d. Length of the operation to repair the rectum.

Answer 1: (a.) and (c.) Continence as the bony development correlates with development of perineal nerves and sphincter muscles. The sacral ratio is a measure of sacral development and is consistent with the development of surrounding musculature and nervous tissues. This ratio can help to predict associated urologic anomalies.

2. In a male patient with an anorectal malformation, the distal colostogram demonstrates a rectoprostatic fistula as shown below



During the dissection of the distal rectum, special care needs to be taken to avoid injury to:

a. The vas deferens
b. An ectopic ureter
c. The seminal vesicles
d. The bladder neck
e. All of the above

Answer 2: (e.) All of the above. With a fistula at this level, the surgeon should take special care to avoid injury to nearby genitourinary structures.

3. What are the indications for the treatment of a rectoperineal fistula in a female?

   a. Relocating the anus to within the sphincter mechanism.
   b. Making the anal opening an appropriate size.

c. Creating an adequate length perineal body.
d. All of the above.

Answer 3: (d.) All of the above. The treatment of a rectoperineal fistula in a female will be indicated by the anal size and location (whether it resides within the sphincter complex) as well as the length of the perineal body.

## Further Reading

Levitt MA, Peña A. Outcomes from the correction of anorectal malformations. Curr Opin Pediatr. 2005;17(3):394–401.

Levitt MA, Peña A. Anorectal malformations. Orphanet J Rare Dis. 2007;2(1):33. https://ojrd.biomedcentral.com/articles/10.1186/1750-1172-2-33.Accessed 1 Nov 2021

Vilanova-Sanchez A, Levitt MA, editors. Pediatric colorectal and pelvic reconstructive surgery. 1st ed. Boca Raton: CRC Press; 2020.

Wood RJ, Levitt MA. Anorectal malformations. Clin Colon Rectal Surg. 2018;31(2):61–70.

# Congential Lung Malformations

**Melissa Suh and Shahab Abdessalam**

**Key Points**

1) Rare but clinically important disorders that can have significant morbidity and mortality.
2) Often identified during prenatal ultrasonography or discovered incidentally.
3) Symptoms range from acute respiratory distress in the newborn to asymptomatic and incidentally discovered.
4) Carry an excellent prognosis in the absence of associated anomalies.

1. Epidemiology:
    a. Incidence: Up to 1 in 2000 live births, detection of asymptomatic lesions increasing with prenatal ultrasonography
    b. Etiology: Abnormal pulmonary embryogenesis
        i. Abnormal budding of the foregut (bronchogenic cysts, extralobar sequestrations)
        ii. Airway obstruction during development (congenital pulmonary airway malformation, intralobar sequestrations, congenital lobar emphysema)
2. Pathophysiology:

    Congenital pulmonary airway malformation (CPAM) (Fig. 1)

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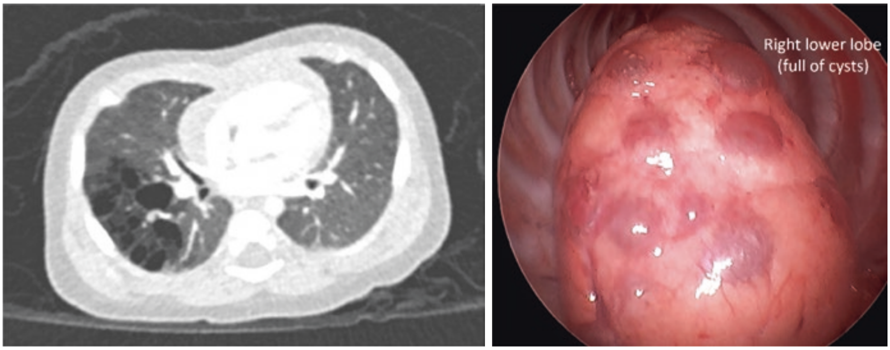

**Fig. 1** Congenital pulmonary airway malformation (CPAM). Computed tomography demonstration and intraoperative view. (Source: Shahab Abdessalam personal file)

a. Anatomy:
   i. Previously known as congenital cystic adenomatoid malformation (CCAM), CPAM is rare but is one of the most common congenital lung abnormalities. It is the second most common cause (25%) of newborn respiratory distress. The most common cause of neonatal respiratory distress is transient tachypnea of the newborn (TTN), responsible for 40% of cases [1]. Other common surgical causes of newborn respiratory distress are congenital diaphragmatic hernia, tracheoesophageal fistula, choanal atresia, pulmonary sequestration, and congenital lobar emphysema [2].
   ii. The lesions are hamartomatous proliferation of cysts composed of adenomatous elements and resemble bronchioles.
   iii. They have connections with the tracheobronchial tree.
   iv. The arterial supply and venous drainage are typically within the pulmonary circulation, but they can have systemic circulation.
   v. Lesions can be multifocal and even bilateral.
   vi. During gestation, compression of heart/central veins by CPAM can lead to hydrops. Compression of GI tract (esophagus) can lead to polyhydramnios.
   vii. Clinical course: when diagnosed prenatally, often born with no symptoms and normal chest radiography. Some prenatally diagnosed CPAM lesions will shrink and disappear on postnatal imaging (5%). Follow-up imaging (CT) is recommended at 3 months if lesion identified prenatally and asymptomatic. If it persists after birth, it is unlikely to resolve.
   viii. Risk of pneumonia: compression of the lesion on bronchial passages leads to overinflation of distal alveoli, trapping of air and mucus, and recurrent pneumonia, which makes future resection potentially more difficult with increased blood loss.

    ix. Risk of neoplasm: if not treated, certain types of CPAM can develop into malignancy such as bronchoalveolar carcinoma, adenocarcinoma, or sarcoma. There is controversy whether pleuropulmonary blastoma develops in CPAMs or whether they are indistinguishable from each other. The primary problem with differentiating pleuropulmonary blastoma from CPAM is that no imaging technology can accurately distinguish these two entities.

b. Classification schemes:

   Based on size and tissue characteristics

    i. Type 0: These lesions arise from the tracheal or bronchial tissue and are the rarest form. The cysts are small and involve the entire lung. Affected infants die at birth due to severely impaired gas exchange.
    ii. Type I: Most common (60–70%). Originate from the distal bronchi or proximal bronchioles and consist of well-differentiated tissue. The cysts are usually single but may be multi-loculated. The majority of cases involve only one lobe of the lung. ~~It has~~ These lesions have malignant potential.
    iii. Type II: This type comprises about 15–20% of cases and consists of multiple cysts that blend into adjacent normal tissue. The lesions resemble dilated terminal bronchioles. Up to 60% of cases have other congenital anomalies. These lesions do not have malignant potential.
    iv. Type III: These lesions comprise about 5–10% of cases and are often very large, proliferating from the distal airways or airspaces and involving an entire lobe or several lobes. This type has not been associated with malignancy.
    v. Type IV: These lesions comprise 10–15% of cases and have a maximum diameter of 7 cm. They are strongly associated with malignancy.

Congenital lobar emphysema (CLE) (Fig. 2)

a. Anatomy:

    i. CLE is massive overinflation of one of the lobes of lung, usually due to airway obstruction from either intrinsic or extrinsic compression.
    ii. Most commonly affects one of the upper lobes of the lung.
    iii. Intrinsic obstruction is more common and often caused by defects in the bronchial wall such as bronchial cartilage deficiency, resulting in airway collapse during expiration. Meconium or mucous plugs, granulomas, or mucosal folds can also cause partial airway obstruction of a lower airway.
    iv. Extrinsic compression may be due to a bronchogenic cyst, obstructing lymph nodes, or vascular anomalies such as a pulmonary artery sling or anomalous pulmonary venous return.
    v. Bronchial atresia has also been identified as a common finding.

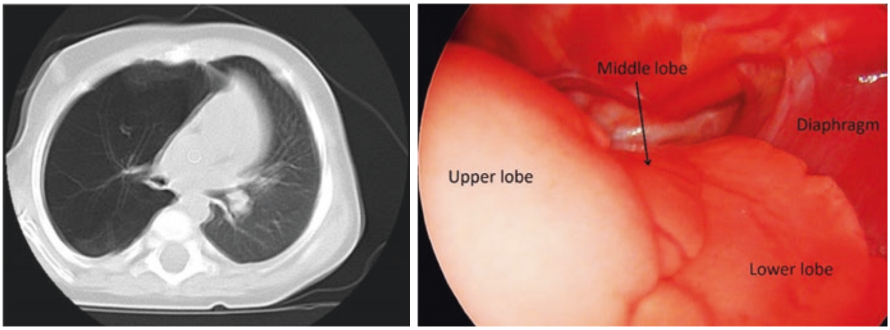

**Fig. 2** Congenital lobar emphysema. Computed tomography and intraoperative photograph. (Source: Shahab Abdessalam personal file)

- vi. Overdistention of one or more lobes of the lung leads to compression of the remaining lung tissue and risk of pulmonary compromise with shift of the mediastinum.
- vii. The conducting airways are typically normal in size and number, but the alveoli within the gas exchange units are enlarged and increased in number.
- viii. There is a 10% risk of associated cardiac abnormality.

Bronchopulmonary sequestration (BPS) (Figs. 3 and 4)

a. Anatomy:

  - i. These are pieces of parenchymal lung separated from the respiratory tree. May be intralobar or extralobar.
  - ii. The blood supply is from systemic circulation. The systemic arterial supply is from an anomalous artery arising from the aorta rather than the pulmonary artery. Sometimes the artery will cross the diaphragm. Intralobar sequestrations drain into the pulmonary vein. Extralobar sequestrations usually drain into the systemic circulation but may drain into the pulmonary vein.
  - iii. Usually occur in the lower lobes.
  - iv. Sequestrations can be attached to or communicate with the GI tract (esophagus or stomach). Referred to as Rokitansky lobe.

b. Classification schemes:

  - v. Intralobar: within the lung parenchyma: (50%) intralobar lesions are found within the normal lung and pleura and almost always in the lower lobes. They occur equally on both sides. Venous drainage is usually through the pulmonary vein.
  - vi. Extralobar: outside of the normal lung: (50%) extralobar lesions are found outside of the pleural space and the venous drainage is usually into

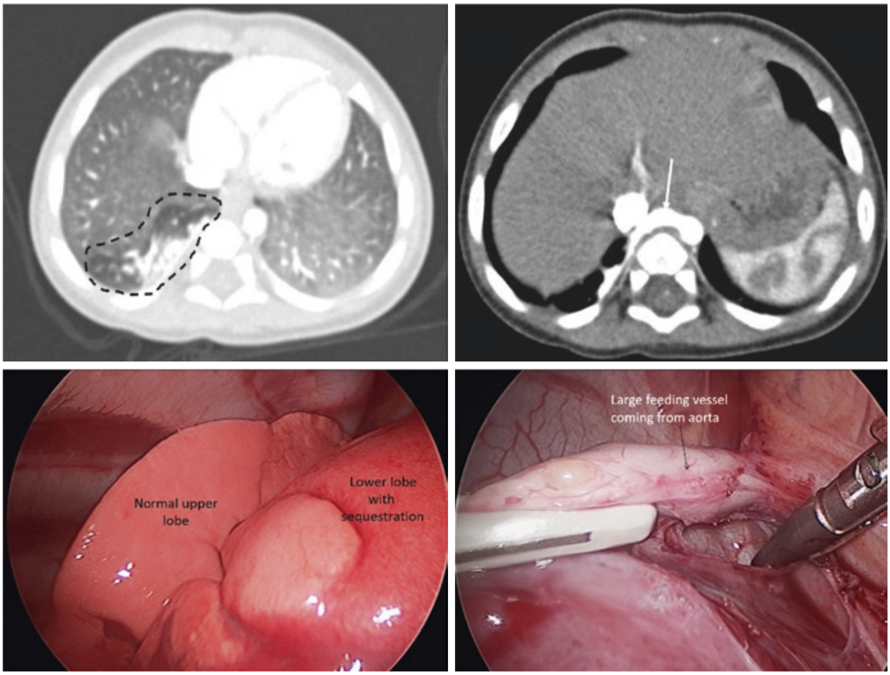
Normal upper lobe
Lower lobe with sequestration
Large feeding vessel coming from aorta

**Fig. 3** Sequestration (intralobar). Aortic blood supply of sequestration is demonstrated on computed tomography of chest. Intraoperative photographs demonstrating aortic blood supply. (Source: Shahab Abdessalam personal file)

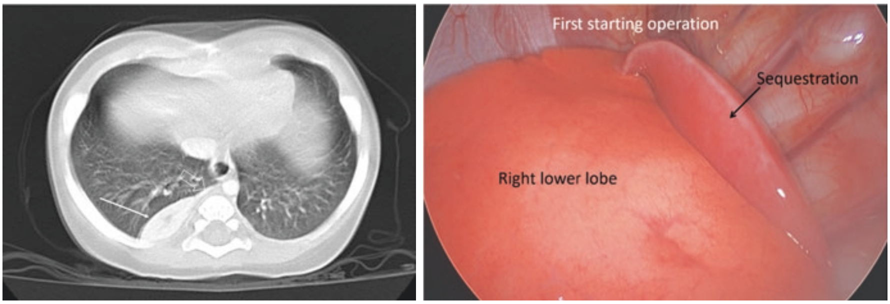
First starting operation
Sequestration
Right lower lobe

**Fig. 4** Sequestration (extralobar). Computed tomography and intraoperative photograph. (Source: Shahab Abdessalam personal file)

the azygos or hemiazygos veins but can be into the pulmonary veins. Two-thirds of the patients may have other congenital abnormalities such as diaphragmatic hernias.

vii. Sequestrations may be transdiaphragmatic sequestrations or entirely intra-abdominal.

Bronchogenic cyst (Fig. 5)

a. Anatomy:
    i. Arise from an abnormal budding or duplication of the ventral foregut tissue present in the airway.
    ii. 15% are intrapulmonary and 85% are mediastinal.
    iii. The cysts may be central or peripheral and may be filled with air, fluid, or both.
    iv. Can cause compression of a bronchus or trachea.
    v. Can result in pulmonary hemorrhage or infection.
    vi. May resemble esophageal duplication as both are foregut structures.
        1. Esophageal duplications will be lined with squamous epithelium and contain both layers of muscularis propria.
        2. Bronchogenic cysts will be lined with cuboid or columnar ciliated epithelium.

3. Clinical features:
    a. Frequently diagnosed prenatally by sonography.
    b. Often asymptomatic and clinically silent at birth.
    c. May cause bronchial compression and hyperinflation.

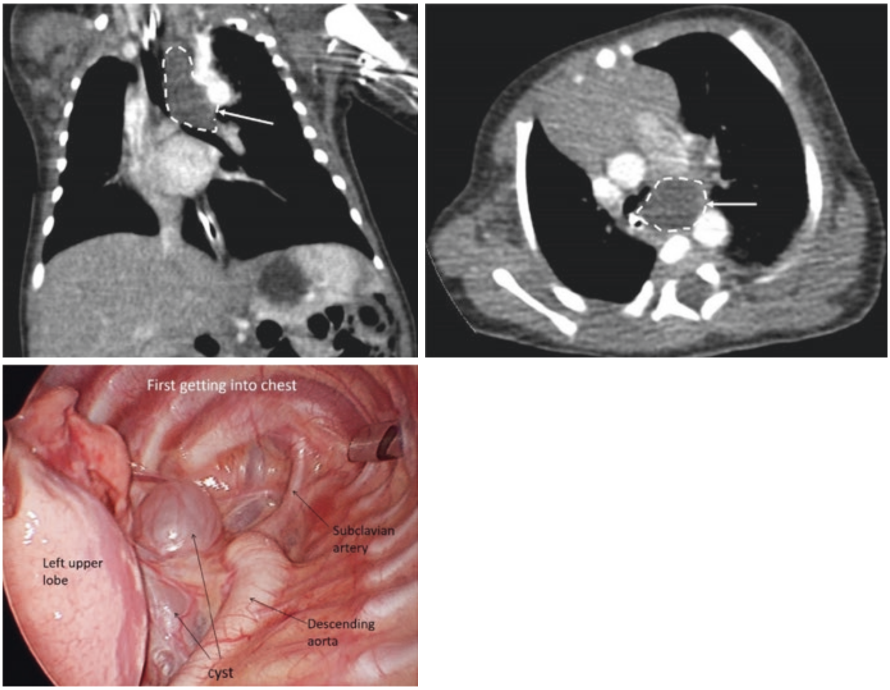

**Fig. 5** Bronchogenic cyst. Demonstrated on computed tomography and intraoperative photograph. (Source: Shahab Abdessalam personal file)

   d. May cause recurring pneumonia.
   e. May present as emergent pulmonary compromise.
   f. Bronchogenic cysts can have drainage into the bronchial tree.

4. Diagnosis:

   a. Symptoms: range from asymptomatic to life threatening.
   b. Radiographic studies:

      i. Prenatal sonography: can diagnose lung lesions as cystic or solid and can track size as the fetus grows.
      ii. Chest radiograph: will reveal fluid-filled structures, such as a bronchogenic cyst, can detect hyperinflation of CLE, and if pneumonia is present.
      iii. Chest sonography: useful in tracking size of CPAM lesions as children grow in utero and can sometimes be used to identify anomalous systemic blood supply of a sequestration.
      iv. Computed tomography (CT) of chest: offers the best delineation of lung lesions and their relationship to normal structures. With intravenous contrast, vascular supply can be identified. Needs to be used sparingly because of the risks of ionizing radiation.
      v. Magnetic resonance imaging of chest: gives good anatomic definition of lesions without ionizing radiation. Requires general anesthesia for small children to remain still for the time necessary for the study. Gadolinium-enhanced studies help identify cystic structures, communication with hollow viscus, and vascular supply.
      vi. Echocardiography: rule out associated cardiac anomaly.

5. Treatment:

   a. CPAM:

      i. Initial measures: presence or absence of hydrops and growth of the lesion determine if interventions are necessary. Steroids given to the mother for babies <28 weeks gestation can stop or potentially reverse growth. If <28 weeks and large cystic component present, then consideration for thoracoamniotic shunt at a center capable of fetal interventions. If no hydrops is present, care is given after delivery.
      ii. Treatment of pneumonia: often a CPAM will be diagnosed while evaluating for recurrent pneumonia. The pneumonia should be treated, and the patient should have a 4- to 6-week period without symptoms before undergoing resection. Resection during active infection can be more difficult and associated with increased blood loss and morbidity. It is a goal of therapy to have the CPAM resected before the child has suffered episodes of pneumonia.
      iii. Period of observation and repeat imaging: some CPAM lesions will resolve spontaneously. When diagnosed prenatally, the presence of symptoms will determine the timing for imaging. If asymptomatic, a

chest X-ray is all that is needed to potentially detect life-threatening abnormalities. CT should be avoided immediately post-delivery as there is considerable artifact. If symptomatic (unable to feed, requiring mechanical ventilation, or inability to wean off oxygen), cross-section imaging should be obtained with either CT or MRI. For those that are asymptomatic, initial cross-sectional imaging should be undertaken at 3–6 months of age. If confirmed to still be present, operative intervention is warranted.

b. CLE:

   i. Initial measures: some patients will have severe ventilatory distress at birth. This occurs when the emphysematous lobe becomes hyperinflated with first breaths and causes mediastinal shift. Chest X-ray may have appearance of a tension pneumothorax. If a chest tube is placed into the emphysematous lung, it can help relieve the pressure, but will create a bronchial leak, making ventilation difficult. If the diagnosis of CLE has been made and a child has severe ventilatory distress, the best course of treatment would be mainstem intubation into the *unaffected* side followed by emergent thoracotomy.
   ii. When a child has mild or no symptoms, it is safe to wait until 6 months of age to see if growth of the child results in relief of the obstruction. If still present after 6 months of age, resection of the emphysematous lobe can be done on an elective basis. Resection of CLE will allow the other lobes in the hemithorax to grow normally as alveolar growth continues to the age of 7–8 years.

c. Bronchopulmonary sequestration:

   i. Timing of the resection is again determined by the presence/absence of symptoms. If symptomatic, then operate. If asymptomatic, then wait until around 6 months of age to operate.
   ii. Preoperative preparation

      1. Care must be taken to identify and ligate the systemic blood supply of the sequestration. When possible, this blood supply is identified preoperatively. These vessels are often short, arise directly from the aorta, and can traverse the diaphragm. When not properly ligated, they will retract and result in significant and potentially fatal bleeding.

   iii. Operative treatment

      1. Thoracoscopic: most intralobar sequestrations will require a lobectomy for complete removal. For those that are multifocal, then consideration for wedge resections (pulmonary sparing). For extralobar sequestrations, simple removal of the sequestration is all that is needed.
      2. Thoracotomy: open resection may be needed when surrounding infection makes removal difficult.

3. Abdominal component: when present, the abdominal component can usually be removed through the diaphragm or behind the diaphragm and is best approached laparoscopically. When a diaphragmatic hernia is also present, it can be repaired after resection of the sequestration. Rarely an additional laparotomy will be needed.

d. Bronchogenic cyst:

    i. Lesions should be resected when detected.
    ii. They can lead to compression of pulmonary, GI, or vascular structures.
    iii. Cysts can become infected and be associated with pneumonia.
    iv. Resection should not be performed during active infection/pneumonia because there is increased inflammation and risk of hemorrhage during procedure.
    v. Thoracoscopic or open approach can be used.

e. Postoperative treatment: standard postoperative care. Decision to place a chest tube should be based upon whether normal lung tissue was entered or a bronchus divided. For most extralobar sequestrations and bronchial cysts, a chest tube can be avoided.

f. Complications to watch for:

    i. Pneumothorax
    ii. Bleeding
    iii. Recurrence
    iv. Malignancy.

## Additional Notes

Congenital lung malformations are frequently benign and asymptomatic. Previous terminology distinguished cystic adenomatoid lesions from bronchopulmonary sequestrations. Recognition that hybrid lesions containing elements of both were common led to terminology which now refers to these lesions as congenital pulmonary adenomatoid malformations. Resection is curative with excellent overall long-term prognosis if there are not additional associated anomalies. Complications including malignancy, infection, torsion, or high-output cardiac failure can present during expectant management. Early elective resection may allow for compensatory lung growth as an infant although the data is mixed. There is ongoing controversy about the need and timing of resection in asymptomatic lesions. Surgery for symptomatic congenital lung malformations have been shown to have longer and more complicated postoperative courses.

Infection is a well-known complication with recurrent pulmonary infection occurring in up to 30% of patients. Prior infectious episodes may increase the risk of bleeding and conversion from thoracoscopy to thoracotomy due to adhesions or inflammation. Subclinical infections may be ongoing and exacerbated by operative manipulation. Asymptomatic patients may have experienced subclinical infections which may complicate operative intervention.

Approximately 4% of malignant lung tumors in children, including adenocarcinoma, squamous cell carcinoma, and rhabdomyosarcoma, are associated with CPAMs. CPAMs have been shown to have an approximately 2% risk of PPB which has a very similar radiographic appearance. It is controversial whether CPAMs give rise to PPB or are indistinguishable on imaging which has made it questionable whether CPAMs give rise to PPBs. Histologic examination is the only way to provide a definitive diagnosis. Some argue that resection may not be fully preventative of malignancy.

Other rare complications have been reported such as high-output cardiac failure associated with lesions with a systemic blood supply, primarily bronchopulmonary sequestrations. Lesions can present with acute pain secondary to torsion around a narrow pedicle with ischemia or acute infarction. Spontaneous regression has been described on prenatal ultrasound in some patients, but the lesion may still be present on CT scan.

If early resection (6–12 months of age) is performed, there is decrease in infection risk and increased likelihood of completing a thoracoscopic operation. This decreases recovery time and functional limitations. Overall, once resected, patients have an excellent prognosis. A suggested time interval of age 6–12 months for intervention to decrease potential operative complications due to small patient size (if done prior to 6 months), decrease chance of recurrent infections (if done later than 1 year), and increase the likelihood of completing a thoracoscopic resection.

## Study Questions

1) Question 1: During a routine prenatal US at 24 weeks gestation, the fetus is found to have a large microcystic CPAM with evidence of mediastinal shift, the best initial management would be:

   a. Immediate delivery of the baby
   b. Thoracoamniotic shunt
   c. Fetal lobectomy
   d. Administration of maternal steroids

Answer to Question 1: “D”. The administration of maternal steroids, especially prior to 28 weeks gestation, has been shown to result in stabilization of the fetus and the lung lesion in 80–95% of cases. Delivering a fetus at 24 weeks gestation, although possible, will result in the need to deal with the sequelae of prematurity in addition to the lung lesion. Thoracoamniotic shunts are most useful when there is a dominant cyst to drain. Given the microcystic findings on this patient, that would not be beneficial. Finally, a fetal lobectomy, while effective in experienced hands at the proper center, poses risks to the mother as well as the potential for preterm delivery and would not be the best first-line option.

2) Question 2: When evaluating an asymptomatic 1-day-old patient with a prenatal diagnosis of a CPAM, proper workup includes:

   a. A CXR
   b. An US of the chest
   c. A CT scan of the chest
   d. An MRI of the chest

Answer to Question 2: “A”. A CXR rules out potentially life-threatening problems such as a pneumothorax or mediastinal shift and is relatively inexpensive and easy to obtain. Although CXR will often not make the diagnosis of the CPAM, in an asymptomatic baby, it is not necessary at 1 day of age. An US is useful in avoiding radiation, but postnatally is suboptimal in delineating anatomic detail and there is considerable motion artifact. Like a CXR, an US should not be used to rule out an abnormality. A CT scan is useful in operative planning, but in a newborn, there is significant lung atelectasis/water that can obscure subtle CPAM findings and given that there is no need for immediate intervention, it is better to wait until 3–6 months of age to obtain the study. This will avoid the need to perform more than one preoperative CT and limit radiation exposure. An MRI not only has the same limitations in the newborn of increased artifact but also has the drawback of needing to be put under general anesthesia because of the length of time needed to perform the study without motion. There is also concern about administering gadolinium to a newborn. For these reasons, the only study necessary in an asymptomatic newborn is a CXR.

3) Question 3: A 3-week-old, former 35-week gestation male has a CXR done for PICC line placement and the radiologist comments that the right upper lobe is enlarged with compression on the lower and middle lobes. The baby is on room air and working on oral feedings with no respiratory distress. A CT scan is performed as well as a bronchoscopy and confirms a narrow but patent right upper lobe bronchus with no external compression. The best recommendation at this point is:

   a. Take the baby to the operating room for an emergent upper lobectomy.
   b. Take the baby to the operating room for an upper lobectomy just prior to discharge.
   c. Continue following CXRs until resolution or 6 months of age as long as he remains asymptomatic.
   d. Reassure the family that no operative intervention will be necessary.

Answer to Question 3: “C”. The case in point is describing congenital lobar emphysema. Management in symptomatic babies includes removal of the affected lobe which may be urgent/emergent depending upon the clinical circumstances. For those that are asymptomatic, such as this patient, observation with serial CXRs (obtained monthly) up until the age of 6 months is preferred. On occasion, the babies will outgrow the malacia/narrowing, but beyond 6 months, this is unlikely. To wait beyond 6 months can lead to further compromise of the unaffected lobes, so intervening at this point is appropriate.

## References

1. Hermansen CL, Lorah KN. Respiratory distress in the newborn. Am Fam Physician. 2007;76(7):987–94.
2. Gallacher DJ, Hart K, Kotecha S. Common respiratory conditions of the newborn. Breath. 2016;12:30–42.

## *Further Reading*

Ben Ameur S, Daoud E, Bouraoui A, et al. Congenital lung malformations: analysis of 27 cases. Tunis Med. 2020;98(6):505–12.

Cortes-Santiago N, Deutsch GH. Pediatric cystic lung lesions: where are we now? Surg Pathol Clin. 2020;13(4):643–55.

Elhattab A, Elsaied A, Wafa T, et al. Jan 27. J Pediatr Surg. 2021;2021

Gulla KM, Parihar MS, Jat KR, Agarwala S, Lodha R, Kabra SK. Congenital lung malformations: experience from a tertiary care center in India. Indian Pediatr. 2021;58(2):129–33.

Lima JS, Camargos PA, Aguiar RA, Campos AS, Aguiar MJ. Pre and perinatal aspects of congenital cystic adenomatoid malformation of the lung. J Matern Fetal Neonatal Med. 2014;27(3):228–32.

Wong MCY, Faure Bardon V, Farmakis K, et al. Ultrasound detected prenatal hyperechoic lung lesions and concordance with postnatal findings: a common aspect for multiple diagnoses. Prenat Diagn. 2021;41(3):323–31.

Zheng J, Tang H, Xu H, Li J, Mao X, Liu G. Thoracoscopic versus open resection for symptomatic congenital pulmonary airway malformations in neonates: a decade-long retrospective study. BMC Pulm Med. 2021;21(1):82.

# Meconium Disorders

**Niti Shahi and Jeremy T. Aidlen**

**Key Points**

1) Meconium disorders of the newborn include distinct entities of meconium ileus, meconium plug syndrome, meconium cyst, and meconium peritonitis.
2) Tenets of nonoperative management for meconium ileus include aggressive IV fluid hydration, NGT decompression, serial water-soluble contrast enemas to evacuate meconium, and N-acetylcysteine therapy.
3) Meconium ileus has over an 80% association with cystic fibrosis (CF). Infants with meconium ileus should undergo genetic testing and sweat test for evaluation for cystic fibrosis.
4) In presence of clinical peritonitis, obstruction, or evidence of perforation, exploratory laparotomy is required. Evacuation of meconium versus enterostomy is recommended.
5) Meconium plug syndrome can be treated by hyperosmolar contrast enema with evacuation of the plugs.
6) Meconium cyst is associated with a contained perforation. Exploratory laparotomy with enterostomy is the treatment.
7) The hallmark of meconium peritonitis is calcification within the peritoneal cavity. Surgery is not always required as the perforation may have occurred in utero and sealed prenatally.

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## Meconium Ileus

1. Pathophysiology
   a. Meconium ileus is caused by obstruction from thickened/inspissated meconium in small bowel, most commonly in terminal ileum [1].
   b. It is associated with cystic fibrosis (up to 15% of cystic fibrosis infants), and cystic fibrosis transmembrane conductance regulator (CFTR) testing should be obtained for patients with suspected meconium ileus [2–4].
2. Incidence
   a. The most common cause of meconium ileus is cystic fibrosis. The abnormally concentrated excretion of epithelial cells results in a dry thick succus. Approximately 80% of infants with meconium ileus have cystic fibrosis [5].
   b. The incidence is 1/1000 births [5].
   c. It is associated with jejunoileal atresia [6]. This may be due to a segmental volvulus in utero of a meconium filled loop of bowel.
   d. Meconium ileus equivalent is a related condition occurring in adolescents with cystic fibrosis in whom succus becomes thickened and difficult to pass resulting in small-bowel obstruction, also known as distal intestinal obstruction syndrome (DIOS) [5].
3. Clinical presentation
   a. Meconium ileus can result in obstruction, perforation, small-bowel volvulus, intestinal atresia, and peritonitis [5].
   b. There are two types of meconium ileus: simple and complicated [6].
      i. Simple: obstruction of terminal ileum with meconium.
      ii. Complicated: meconium pseudocyst, atresia, perforation, or volvulus.
   c. Symptoms include failure to pass meconium, difficulty feeding, abdominal distension, and bilious emesis 1–2 days after birth [4–6].
   d. Physical examination will reveal a neonate with softly distended abdomen, sometimes with palpable loops of intestine [5]. Complicated patients may have a more concerning exam with features including abdominal wall erythema, edema, or tense abdominal distention with or without tenderness.
   e. NGT output is generally bilious.
4. Diagnosis
   a. *Ultrasound*: Ultrasound demonstrates a hyperechoic mass (inspissated meconium) with dilated bowel [4, 7] (normally, stool is hypoechoic) [6].
   b. *X-ray*: There are no air-fluid levels because of viscous meconium. X-ray demonstrates dilated bowel loops and "soap bubble" or "ground glass opacities" [6]. Often, distal bowel and colon are not visible.
   c. *Water-soluble contrast enema*: Water-soluble contrast enemas are diagnostic and therapeutic; they may also assist with evaluation for malrotation, atresia,

and microcolon [6]. If contrast does not reflux into terminal ileum, one may need to consider repeat attempts or surgical management [6].

    i. Barium is avoided for two reasons. First, if there is a perforation and leak, extravasation of barium will cause peritonitis. Second, barium will thicken in the lumen of the colon and can contribute to the obstruction [5].

        1. Contrast enema will show thick plugs and ropes of material occluding the colon and distal bowel. Irrigation of the colon with the water-soluble contrast can help the patient evacuate these plugs [5].

  d. Evaluation for cystic fibrosis.

    i. Initial evaluation with sweat testing is recommended for cystic fibrosis; Chloride >60 mmol/L in sweat is diagnostic [6]. CF can be diagnosed by either Na+ or a Cl− level above 60 mmol/L, although the typically used criteria is chloride level.
    ii. Confirmatory CFTR testing should follow sweat testing. The CFTR gene is autosomal recessive and is on chromosome 7q21.

5. Treatment: (Fig. 1)

  a. Hydration is essential as there is a high risk of intravascular losses with meconium ileus [8]. Resuscitation with IV fluids ≥150 cc/kg/day is recommended for a full-term infant. These patients also require correction of electrolyte abnormalities [6].
  b. Bowel rest and nasogastric decompression is recommended [6].
  c. One percent N-acetylcysteine (high osmolality) or gastrograffin (low osmolality) solution, saline, or glycerin enemas can be performed to help break up the meconium [6].

    i. Up to two attempts are recommended [6].
    ii. One can consider administration of N-acetylcysteine via NGT after enemas 5 cc every 6 h [6].
    iii. There is a risk of iatrogenic perforation with enemas [8].
    iv. Many providers have described use of higher concentrations of NAC (10%) for antegrade or retrograde treatment of meconium ileus.

  d. The majority of cases of meconium ileus are successfully managed nonoperatively [8].
  e. Given high incidence of cystic fibrosis, one needs to consider nutritional supplementation with pancreatic enzymes and fat-soluble vitamins A, D, E, and K [8, 9].
  f. Surgical management indicated for worsening clinical status, peritonitis, iatrogenic perforation, and persistent abdominal distension despite nonoperative therapy [6].

    i. Laparotomy can be performed via right-sided supraumbilical transverse incision [1].

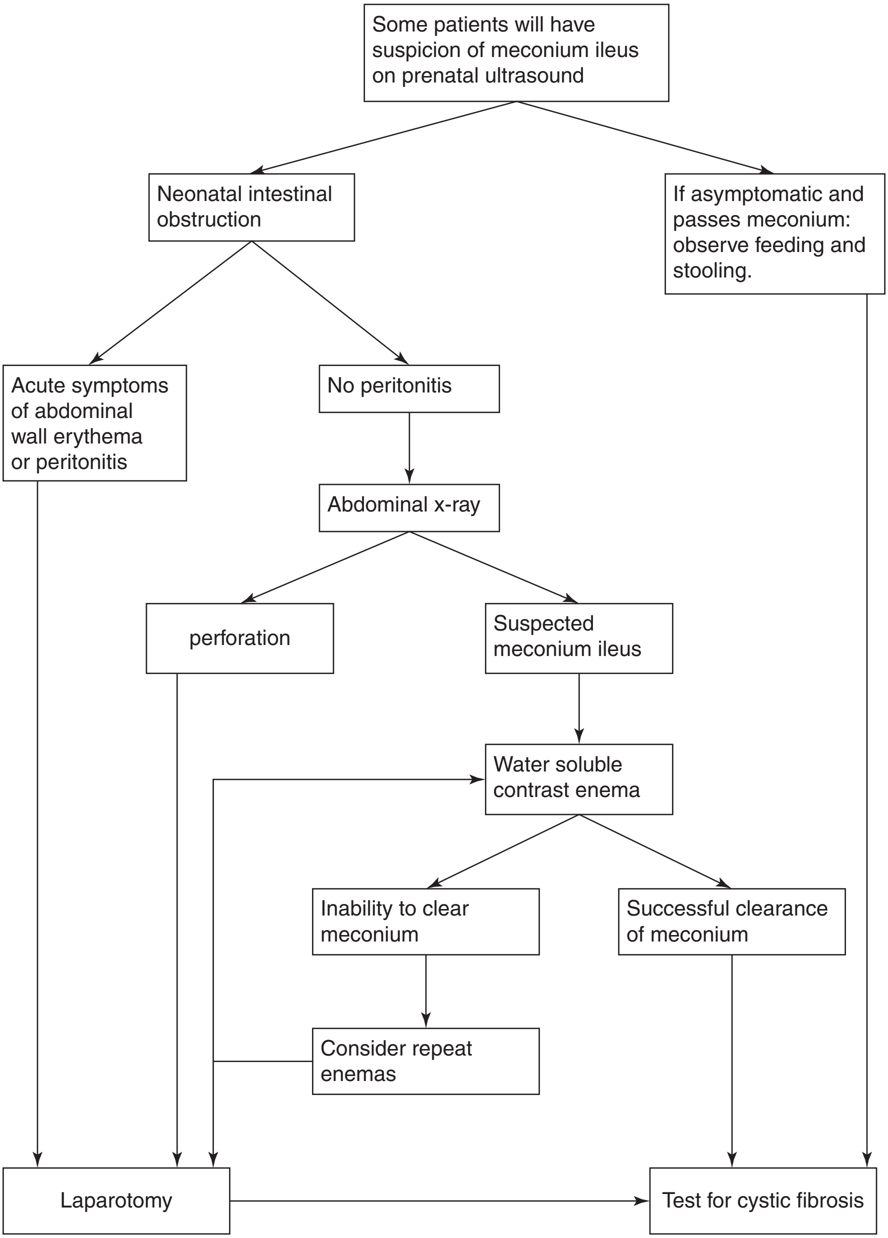
Some patients will have suspicion of meconium ileus on prenatal ultrasound
Neonatal intestinal obstruction
If asymptomatic and passes meconium: observe feeding and stooling.
Acute symptoms of abdominal wall erythema or peritonitis
No peritonitis
Abdominal x-ray
perforation
Suspected meconium ileus
Water soluble contrast enema
Inability to clear meconium
Successful clearance of meconium
Consider repeat enemas
Laparotomy
Test for cystic fibrosis

**Fig. 1** Algorithm for treatment of meconium ileus. (Source: Adapted from: Luks FI, Dolgin S, Berman L, Wagner AJ. Meconium Ileus, in: Hirschl, Ron, et al., eds. Pediatric Surgery NaT, American Pediatric Surgical Association, 2020. www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829040/all/Meconium_Ileus. Downloaded 1 Oct 2021)

   ii. Preferred treatment is evacuation of meconium via enterotomy with closure (without enterostomy). An irrigation tube may be placed as a tube enterostomy.
   iii. Enterostomy options include proximal ileostomy with distal mucous fistula (most common), Mikulicz double-barreled enterostomy, the Bishop–Koop distal chimney enterostomy (end to side anastomosis), the Santulli and Blanc proximal enterostomy (larger stoma; can be used if short mesentery), and tube enterostomy (often T tube) [1, 6] (Fig. 2).
   iv. Irrigations may continue via stoma anterograde or retrograde [8].
   v. Irrigation and suction better than milking meconium as it is typically gentler [1]. This can be done via an enterotomy site and occasionally with a small tube placed through an intentional appendicostomy (Malone).

6. Postoperative complications

   a. Complications include recurrent or persistent meconium obstruction (most common), failure to thrive, pancreatic insufficiency, short bowel syndrome, bowel injury/enterotomy, pulmonary complications, adhesive bowel obstructions, and malnutrition (especially from fat-soluble vitamin deficiency).

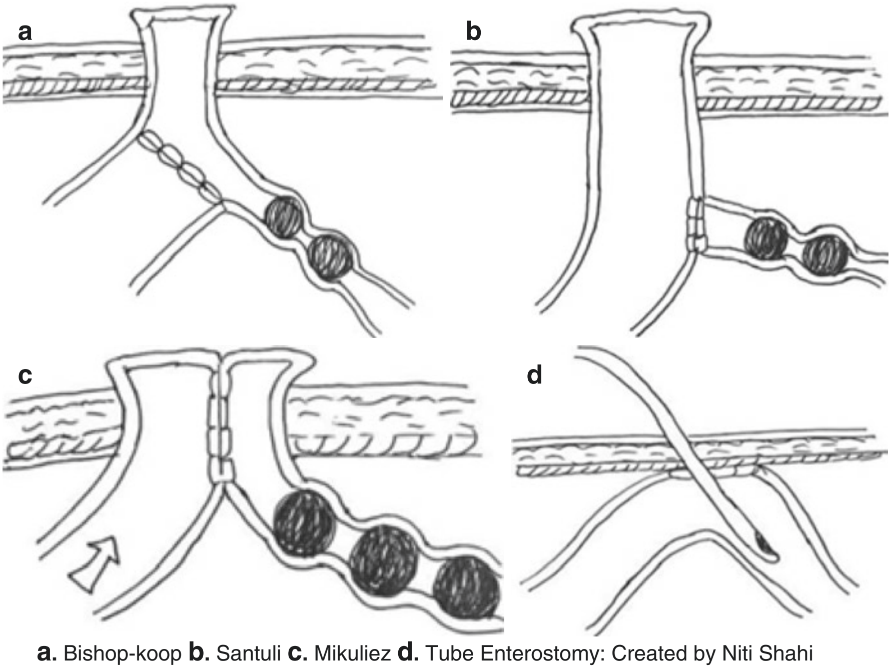
a
b
c
d
**a.** Bishop-koop **b.** Santuli **c.** Mikuliez **d.** Tube Enterostomy: Created by Niti Shahi

**Fig. 2** Enterostomy options available in the treatment of meconium disorders. (Source: Niti Shahi, MD)

## Meconium Plug Syndrome

1. Pathophysiology
   1. Meconium plug syndrome is caused by either obstruction from inspissated meconium in the colon or colonic dysmotility [6, 10].
   2. Other terms for meconium plug syndrome are colonic immaturity and functional colonic obstruction [5].
   3. Meconium plug syndrome can be associated with Hirschsprung disease in approximately 40% of cases, and it is associated with cystic fibrosis in approximately 40% of cases [5].
   4. Meconium plug syndrome is often encountered in very low–birth-weight infants [9].
2. Incidence
   1. The incidence is 1/3000 births [5].
3. Clinical presentation
   1. Meconium plug syndrome has similar symptoms but is considered more benign than meconium ileus [6].
   2. Plugs appear dark green with pale head [8].
   3. Patients generally have improvement of symptoms after passage of plugs unless the underlying diagnosis is related to dysmotility or Hirschsprung disease [5].
4. Diagnosis
   1. *Water-soluble contrast enema*: Water-soluble contrast enemas are diagnostic and therapeutic (similar to meconium ileus).
   2. Some patients with meconium plug syndrome will have Hirschsprung disease which is diagnosed by absence of ganglion cells on a rectal biopsy [5]. Enemas do not distinguish between these disorders effectively as both will have a transition zone with proximal bowel dilation.
   3. One should consider evaluation of thyroid hormone levels as meconium plug syndrome may be associated with congenital hypothyroidism [6].
5. Treatment
   1. The treatment is the same as meconium ileus.
   2. Meconium plug syndrome is usually managed nonoperatively and often spontaneously resolves [8].

## Small Left Colon Syndrome

1. Clinical presentation: The presentation of small left colon syndrome is identical to meconium plug syndrome.
    a. Small left colon syndrome is more common in infants of diabetic mothers.
    b. It may be related to impaired glucagon metabolism.
2. Diagnosis
    a. Water-soluble contrast enema: Water-soluble contrast enemas can be diagnostic and therapeutic and demonstrates microcolon of left colon only with more proximal meconium plugs and distention.
3. Treatment
    a. Enema is typically therapeutic, and the patient can be fed after meconium evacuation.
    b. This is typically a self-limited condition. If symptoms persist, consider Hirschsprung's disease as the presentation and enema findings are similar.

## Meconium Peritonitis

1. Pathophysiology
    a. Meconium peritonitis can be caused by volvulus or perforation of meconium pseudocyst or perforation from meconium ileus in utero [4, 7, 8].
2. Incidence
    a. The incidence is 1/30,000 births–1/35,000 births [11, 12].
3. Clinical presentation
    a. Symptoms include vomiting, abdominal distension, and respiratory distress [13].
4. Diagnosis
    a. *Ultrasound*: Ultrasound demonstrates free fluid, speckled calcifications [6].
    b. *X-ray*: X-ray demonstrates speckled "eggshell" calcifications [6].
5. Treatment
    a. Bowel obstruction requires exploratory laparotomy with irrigation, debridement, ostomy creation, and possible bowel resection [6, 13].
    b. The initial operation is followed by a planned second operation for intestinal re-anastomosis once patient has stabilized [13].

  c. No role for therapeutic enemas in these patients [8].
  d. If volvulus is causative, untwist the bowel and examine after reduction; only necrotic bowel should be removed. Questionable viability should prompt closure and "second look." The abdominal cavity should be irrigated [1].
  e. If meconium peritonitis is incidentally discovered on imaging and clinically well, the patient does not necessarily need operative intervention [8]. This likely represents in utero perforation that has spontaneously healed.
  f. If iatrogenic perforation from enema used to treat meconium obstruction, the patient requires urgent surgical intervention [8].

## Meconium Pseudocyst

1. Pathophysiology

   a. Small-bowel perforation from meconium disorders causes formation of meconium pseudocyst; the perforation typically occurs in utero [6].
   b. Meconium pseudocyst leads to development of a sterile peritonitis [8].
   c. There are four types of meconium pseudocysts: adhesive meconium peritonitis, giant cystic meconium peritonitis or pseudocyst, meconium ascites, and infected meconium peritonitis [6, 14, 15].

2. Incidence

   a. Meconium pseudocysts are rare but are associated with up to 50% of cases of meconium peritonitis [16].

3. Clinical presentation

   a. Associated symptoms include bilious vomiting, abdominal distension, and respiratory distress [13].

4. Diagnosis

   a. *Ultrasound*: Ultrasound demonstrates intra-abdominal calcifications and evidence of cyst with an echogenic rim [6, 17].
   b. *Abdominal X-ray*: Large centrally located air-fluid level.

5. Treatment

   a. Operative management for obstructed patients consists of an exploratory laparotomy, resection of pseudocyst, irrigation, intestinal resection as indicated, and primary anastomosis if feasible [1, 13]. The bowel is located posterior to the pseudocyst. The pseudocyst wall may be adherent to the liver in which case it should not be resected due to risk of bleeding.

      i. Do not need to remove the rind as it risks enterotomy [1].

## Additional Considerations

1. Workup for neonatal obstruction: Consider workup if infant fails to pass meconium within 24–48 h.
    a. *Contrast enema*: Contrast enema should be performed if Hirschsprung disease is suspected. Contrast enema may identify transition zone associated with Hirschsprung disease, microcolon, meconium plug, or meconium obstruction (meconium ileus).
    b. *Suction rectal biopsy*: Following contrast enema, suction rectal biopsy should be performed in a child with small left colon, meconium plug, or delayed passage of meconium to exclude Hirschsprung disease [5].
    c. *Upper GI series*: Malrotation and midgut volvulus should be considered and ruled out if a possibility because missing this diagnosis can lead to a lethal loss of intestine. Malrotation is ruled out with upper gastrointestinal series if suspected. If detected, urgent laparotomy or laparoscopy is indicated [5].

## Additional Notes

Newborns with a meconium disorder are diverse, but each patient can be managed using a standard approach. An orogastric or nasogastric tube should be placed for decompression and intravenous fluid administered to infants with abdominal distention and a failure to pass meconium. Bilious emesis or tube effluent should prompt an upper GI contrast evaluation to rule out malrotation with or without midgut volvulus. If the upper GI series is normal, in the stable, and clinically obstructed newborn, it is usually reasonable to proceed next with a contrast enema.

Frequently, there are early clues that the newborn bowel obstruction is more distal such as multiple distended loops rather than a few on a plain film, but this is not a guarantee that a volvulus is not present, so upper GI contrast should still be obtained prior to a contrast enema in most cases. Additional clues for meconium disorder as the cause of obstruction include intraperitoneal calcifications on plain film or a family history of cystic fibrosis.

The contrast enema is typically diagnostic if not therapeutic in the case of meconium obstruction. In order for the enema to be successfully therapeutic, it needs to traverse the transition zone into dilated proximally obstructed intestinal loops. Inability to fill dilated loops with contrast in a retrograde fashion does not eliminate the diagnosis of intestinal atresia or segmental volvulus, and it should not lead to the conclusion that the obstruction could be relieved nonoperatively. Persistence is crucial, as is effective communication with Neonatology and Radiology teams. Enemas may need to be repeated on multiple occasions.

There remains some debate as to whether every baby who receives a diagnosis of meconium obstruction by contrast enema deserves a suction rectal biopsy to evaluate for Hirschsprung disease. Enemas can look identical in these conditions, though other clinical characteristics may be quite dissimilar. A thorough and thoughtful multidisciplinary approach dedicated to the care of these infants is warranted to facilitate early diagnosis, treatment, and the institution of enteral nutrition. Genetic screening and sweat testing are routine for those diagnosed with meconium disorder. Family counseling and referral to a specialist team is indicated if a diagnosis of cystic fibrosis is secured.

## Study Questions

1) Question 1: Which of the following obstructive conditions is primarily managed nonoperatively?

   a. Meconium Ileus
   b. Meconium pseudocyst
   c. Meconium peritonitis
   d. Hirschsprung disease

Answer to Question 1: (a) Meconium ileus can be diagnosed and successfully treated by enema either with water-soluble contrast such as Gastrograffin or with N-acetylcysteine. The other conditions listed require surgical therapy when obstruction is present.

2) In patients who require operative management for meconium ileus, what is the most common postoperative complication?

   a. Recurrent obstruction
   b. Short bowel syndrome
   c. Dehydration
   d. Pneumonia

Answer to Question 2: (a) Recurrent obstruction, short bowel syndrome, and dehydration are all complications that can be seen in infants with meconium ileus. However, recurrent obstruction either due to adhesions or meconium ileus equivalent is the most common of these complications.

3) A newborn infant fails to pass meconium within the first 48 h of life. An X-ray is obtained demonstrating dilated loops of small bowel. The infant has a distended abdomen and evidence of a patent anus on examination. An NGT is placed with return of bilious output. What is the next best step?

   a. Contrast enema
   b. Upper GI series
   c. Suction rectal biopsy
   d. CFTR testing

Answer to Question 3: (b) Infants who have failed to pass meconium at 48 h are at risk for a variety of disorders including intestinal atresia, malrotation, meconium disorders, imperforate anus, and Hirschsprung disease. Initial evaluation should include hydration and bowel decompression with a NG or OG tube. In patients who have bilious output, an upper GI series should be obtained to rule out malrotation and midgut volvulus. If there is no evidence of malrotation and/or midgut volvulus on UGI series, a contrast enema should be obtained to help evaluate for possible intestinal atresia, meconium disorders, and/or Hirschsprung disease. In infants with a high level of suspicion for Hirschsprung disease, a suction rectal biopsy may also be obtained.

## References

1. Meconium Ileus Procedures [Internet]. American Pediatric Surgical Association and Unbound Medicine, Inc., 2020 [updated May 3, 2016; cited May 10th, 2021]. https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829174/all/Meconium_Ileus_Procedures
2. Bagolan P, Morini F, Conforti A. Meconium ileus. In: Pediatric surgery: general principles and newborn surgery; 2020. p. 973–92.
3. Waldhausen JHT, Richards M. Meconium ileus. Clin Colon Rectal Surg. 2018;31(2):121–6.
4. O'Sullivan BP, Freedman SD. Cystic fibrosis. Lancet. 2009;373(9678):1891–904.
5. Coppola CP, Kennedy AP Jr, Scorpio RJ. Pediatric surgery. Springer; 2016.
6. Holcomb GW III, Patrick Murphy SJ, Peter SD. Holcomb and Ashcraft's pediatric surgery. Elsevier, London; 2020. p. 211–89.
7. Carlyle BE, Borowitz DS, Glick PL. A review of pathophysiology and management of fetuses and neonates with meconium ileus for the pediatric surgeon. J Pediatr Surg. 2012;47(4):772–81.
8. Meconium Ileus [Internet]. American Pediatric Surgical Association and Unbound Medicine, Inc., 2020 [updated November 2, 2020; cited May 10th, 2021]. https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829040/all/Meconium_Ileus
9. Hajivassiliou C. Intestinal obstruction in neonatal/pediatric surgery. Seminars in pediatric surgery. Elsevier, 2003.
10. Cuenca AG, Ali AS, Kays DW, Islam S. "Pulling the plug"—management of meconium plug syndrome in neonates. J Surg Res. 2012;175(2):e43–6.
11. Nam SH, Kim SC, Kim DY, Kim AR, Kim KS, Pi SY, et al. Experience with meconium peritonitis. J Pediatr Surg. 2007;42(11):1822–5.
12. Paladini D, Volpe P. Ultrasound of congenital fetal anomalies: differential diagnosis and prognostic indicators. CRC press; 2018.
13. Minato M, Okada T, Miyagi H, Honda S, Takazawa K, Kubota KC, et al. Meconium pseudocyst with particular pathologic findings: a case report and review of the literature. J Pediatr Surg. 2012;47(4):e9–e12.
14. Dirkes K, Crombleholme TM, Craigo SD, Latchaw LA, Jacir NN, Harris BH, et al. The natural history of meconium peritonitis diagnosed in utero. J Pediatr Surg. 1995;30(7):979–82.
15. Ping LM, Rajadurai VS, Saffari SE, Chandran S. Meconium peritonitis: correlation of antenatal diagnosis and postnatal outcome - an institutional experience over 10 years. Fetal Diagn Ther. 2017;42(1):57–62.
16. Siegel MJ. Pediatric sonography. Lippincott Williams & Wilkins; 2011.
17. Rumack CM, Levine D. Diagnostic ultrasound E-book. Elsevier Health Sciences; 2017.

# Biliary Atresia

**Yi-Horng Lee**

**Key Points**

1) Biliary atresia is a severe progressive condition of biliary fibrosis that must be differentiated from other causes of neonatal jaundice.
2) Biliary atresia is treated with the Kasai hepatic portoenterostomy in which the extrahepatic biliary tree is resected and drainage of bile from the liver is established with an anastomosis between the porta hepatis and a loop of jejunum.
3) Without Kasai portoenterostomy by age 2 months old, there is very little chance of a child establishing normal bile drainage with their native liver, and liver transplantation is the best chance for success.

Biliary atresia is a progressive fibrosing process of the extrahepatic biliary tree in the neonate. It presents with jaundice and is life threatening if flow of bile is not restored. Most infants with biliary atresia are born at full term with normal birth weight. They initially thrive and seem healthy. Scleral icterus and pale colored stool are the most common presenting symptoms, and they can progress to life-threatening hepatic failure if untreated. Timely diagnosis and treatment are paramount to achieving a positive outcome for these infants. Biliary atresia is the most common indication for liver transplantation in children.

1. Clinical presentation
    a. History

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        i. Most are born at term with normal birth weight.
        ii. They initially thrive and seem healthy.
        iii. Scleral icterus and jaundice are initial symptoms.
        iv. Dark urine.
        v. Pale, white, gray, or acholic stools.
        vi. Poor weight gain.
        vii. Itching and irritability.

    b. Physical examination

        i. Scleral icterus
        ii. Jaundice
        iii. Hepatomegaly/splenomegaly

2. Pathophysiology

    a. Incidence

        i. One in 10,000–18,000 births
        ii. Higher incidence in females
        iii. Higher incidence in Asian and Black children

    b. Progressive fibrosis and obstruction of the extrahepatic biliary tree ascending to intrahepatic fibrosis and obstruction are noted.
    c. There is no clearly identifiable etiology; genetic, infectious, toxic, and immunologic factors have been implicated.
    d. Types of biliary atresia

        i. Perinatal biliary atresia (70–85%): isolated biliary atresia without any other anomalies.
        ii. Biliary atresia associated with laterality malformation (10–15%): also known as biliary atresia splenic malformation (BASM). The laterality malformations include situs inversus, asplenia or polysplenia, malrotation, interrupted inferior vena cava and cardiac anomalies.
        iii. Biliary atresia in association with other congenital malformations (5–10%): associated congenital anomalies include intestinal atresia, anorectal malformations, renal anomalies, and cardiac malformations.

    e. Anatomical classifications of biliary atresia differentiate the level of biliary tree involvement.

        i. Kasai classification (Japanese and Anglo-Saxon classification) [1] (Fig. 1)

            1. Type I: Obliterated common bile duct
            2. Type II: Obliterated common hepatic duct

                a. Type IIa: "cystic biliary atresia"-sometimes has a cyst at hepatic hilum. Cystic duct and common bile duct are patent.
                b. Type IIb: Obliteration of common bile duct, common hepatic duct, and cystic duct.

            3. Type III: Obliterated left and right hepatic ducts (90% of cases)

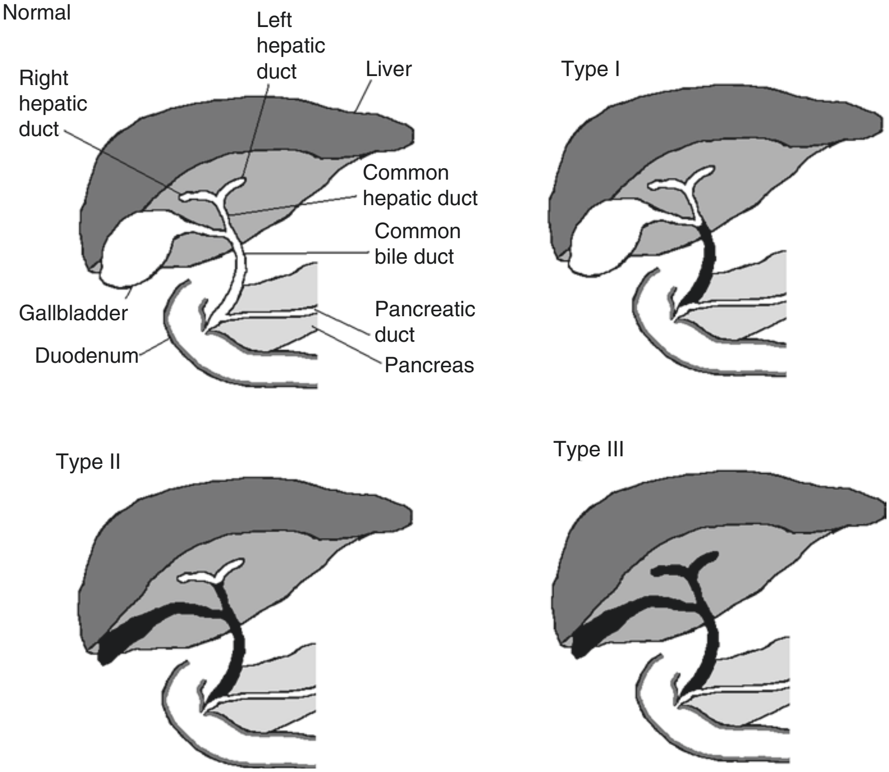
Normal
Left hepatic duct
Right hepatic duct
Liver
Common hepatic duct
Common bile duct
Gallbladder
Duodenum
Pancreatic duct
Pancreas
Type I
Type II
Type III

**Fig. 1** Kasai classification of biliary atresia. Normal anatomy in upper left panel. Type I: Obliterated common bile duct. Type II: Obliterated common hepatic duct. Type III: Obliterated right and left hepatic ducts. (Source: Bryan Walters and Christopher Coppola)

  ii. French classification [2].
    1. Type 1 (3%) Common bile duct obliteration
    2. Type 2 (6%) Obliteration to level of hepatic duct and cyst in liver hilum
    3. Type 3 (19%) Obliteration to level of porta hepatis, patent common bile duct, cystic duct, and gallbladder
    4. Type 4 (72%) Complete extrahepatic biliary tract obliteration

f. Differential diagnosis
  i. Physiologic jaundice of newborn: should clear by 2 weeks of age
  ii. ToRCH infection
    1. Toxoplasmosis
    2. Rubella
    3. Cytomegalovirus
    4. Herpes simplex virus

    iii. Alagille syndrome: paucity of interlobar bile ducts
        1. JAG1 gene mutation is present in more than 90% of cases of Alagille's syndrome.
        2. NOTCH2 gene mutation is present in a smaller number of patients.
        3. Either of these mutations can lead to abnormal signaling during development and result in defects of the bile ducts, facial features, spinal column, and heart [3].
    iv. Choledochal cyst and Caroli disease (intrahepatic ductal dilation)
    v. Alpha 1 anti-trypsin deficiency
    vi. Cystic fibrosis

3. Diagnosis
    a. Laboratory workup
        i. CBC
        ii. LFTs: direct bilirubin and GGTP are preferentially elevated in biliary atresia patients.
        iii. ToRCH infection titers
        iv. Alpha 1 anti-trypsin level
        v. Newborn screen
        vi. Sweat chloride: can be done in infants as early as 2 days of age, but definitely by 10 days of age.
    b. Imaging
        i. Abdominal sonography can show presence/absence of gallbladder, dilation of intrahepatic and extrahepatic bile ducts. The "triangular cord sign" is highly suggestive of biliary atresia. It is important to note that presence of gallbladder does not rule out biliary atresia.
        ii. Hepatobiliary iminodiacetic acid (HIDA) scan should be performed after preparing the patient with 5 days of phenobarbital (5 mg/kg/day) to increase its specificity. Visualization of radioactive tracer excretion into the intestinal loops rules out biliary atresia. Abnormal HIDA may suggest other neonatal hepatic diseases and mandates a liver biopsy and/or cholangiogram. Delayed or attenuated excretion does not definitively exclude biliary atresia and still warrants further evaluation. A normal HIDA scan in a very young infant (i.e., <6 weeks old) may warrant repeat HIDA if cholestasis persists.
        iii. MR cholangiography can effectively rule out biliary atresia if patent biliary tree is demonstrated.
    c. Liver biopsy can diagnose the cholestasis of biliary atresia and identify alternate causes such as hepatitis or paucity of interlobar bile ducts (Alagille syndrome). Findings consistent with biliary atresia include expanded portal tracts with bile duct proliferation, portal tract edema, fibrosis, and

inflammation, and canalicular and bile duct bile plugs. These findings can be seen in other types of biliary obstruction such as choledochal cyst. Hence, cholangiogram is an important adjunct to the diagnosis of biliary atresia. Liver biopsy can be done via open, laparoscopic, or percutaneous approaches depending on local expertise available.

d. Intraoperative cholangiogram is necessary when there is persistent cholestasis due to direct hyperbilirubinemia in an infant who does not have a clearly identifiable cause. This is performed by inserting a small catheter into the biliary tree via the gallbladder, cystic duct, or even common hepatic duct. Water-soluble contrast material is injected into the biliary tree after the common bile duct is dissected out and occluded. Biliary atresia is diagnosed when there is no enhancement of the proximal biliary tree.

e. Few centers have reported the use of neonatal endoscopic retrograde cholangiopancreatography (ERCP) to determine patency of the biliary tree; this is not a common practice.

4. Treatment

   a. Preoperative

      i. Phenobarbital
      ii. Ursodeoxycholic acid
      iii. Water-soluble vitamins

   b. Operative

      i. Exploratory laparotomy for liver biopsy and cholangiogram; must work closely with pathologist
      ii. Kasai procedure (hepatoportoenterostomy)

         1. The extrahepatic biliary tree (gallbladder, common bile duct, and common hepatic duct) is resected.
         2. A Roux limb of jejunum is brought up to the liver hilum in a retrocolic manner, and a downstream jejunojejunostomy anastomosis is created.
         3. An anastomosis is formed between the Roux limb of jejunum and the hilum of the liver. The level of anastomosis at the liver hilum is determined by how extensive the bile ducts have been scarred. Usually, the common hepatic duct is resected, and an anastomosis is made at the "hilar plate," the confluence between right and left hepatic ducts. Patent ducts are usually not visualized, and the bile flow is seen by observing a focal area of intense yellow drainage when placing a piece of gauze adjacent the hilar plate prior to the anastomosis (Fig. 2).

      iii. Frozen section evaluation of a biopsy from the hilar plate can help by revealing that patent biliary ductules have been reached, but this does not always correlate with successfully postoperative bile drainage.

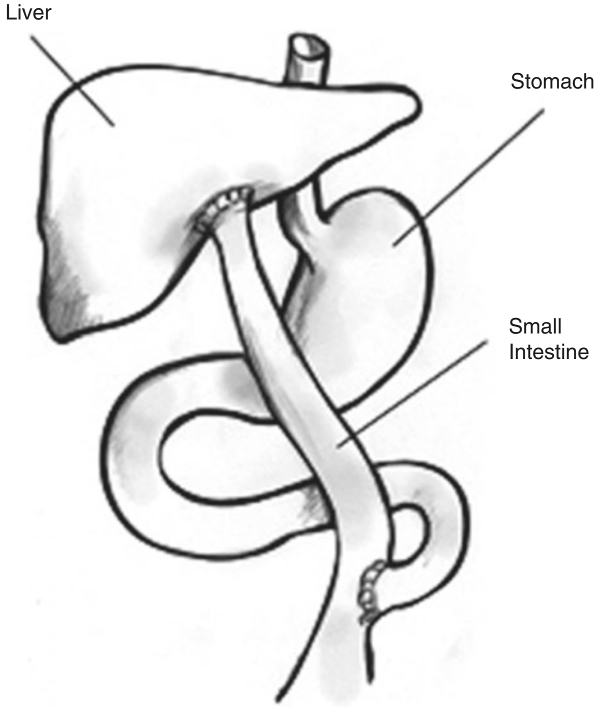

**Fig. 2** Kasai procedure for biliary atresia: hepatoportoenterostomy (Adapted from National Digestive Diseases Clearinghouse, National Institutes of Health, NIH Publication 12–5289, July 2012. http://digestive.niddk.nih.gov/ddiseases/pubs/atresia/#13. Downloaded on 22 Dec 2013)

c. Postoperative care

   i. Patients are maintained NPO with IV fluid until GI function returns.
   ii. Serial LFTs are measured to track drainage of bile and restoration of liver function.
   iii. Postoperative antibiotics are used to reduce the incidence of cholangitis; piperacillin/tazobactam is used perioperatively, and prophylactic dosing of sulfamethozaxole/trimethoprim is used for 3–6 months thereafter.
   iv. Steroids had been used in the past, but there is no benefit shown in a randomized, placebo-controlled trial. Its routine use is no longer recommended.
   v. Ursodeoxycholic acid (15–30 mg/kg/day) aids bile flow.
   vi. Nutritional support is important since there is malabsorption due to cholestasis. We usually need to increase caloric density of the formula given up to 27 kcal/kg in order to achieve 130–150% of the recommended energy intake. There is also a higher-than-expected protein need of 3–4 g/kg/day. Medium-chain triglyceride (MCT) oil is often used.
   vii. Fat-soluble vitamin supplements.

  d. Liver transplantation: when Kasai has not been performed by 3 months of age, or if Kasai procedure is unsuccessful in producing bile drainage from liver, the child needs to be evaluated for liver transplantation.

5. Outcome

   a. Without operation to restore biliary drainage, 50% of children die by age 5 years old.
   b. Complications of Kasai procedure:

      i. Failure to drain bile/jaundice. Bile may drain initially and then cease when the process of atresia extends further into the liver.
      ii. Bile leak
      iii. Cholangitis and sepsis
      iv. Leak from Roux-en-Y anastomosis
      v. Ascites
      vi. Bowel obstruction
      vii. Portal hypertension and formation of varices
      viii. Cirrhosis and liver failure
      ix. Poor growth and malnutrition
      x. Deficiency of fat-soluble vitamins A, D, E, and K

   c. Patients treated with Kasai portoenterostomy before age 2 months old have better prognosis than those treated later.

      i. One-third patients have bile drainage with Kasai.
      ii. One-third will survive with a liver transplant.
      iii. One-third will die.

## Additional Notes

Biliary atresia remains a challenging and life-threatening condition with a very poor prognosis if the child is not able to achieve bile outflow or undergo liver transplantation. Experience has shown that chances of successful bile excretion from the child's native liver are much greater if Kasai portoenterostomy is performed before age 2 months old. A recent report described how better results were achieved in children who had the cystic type of biliary atresia which may indicate it is a distinct entity with a lesser degree of hepatic fibrosis [4].

Patients who undergo a Kasai procedure and establish bile flow still will need close follow-up. Cholangitis and continuation of hepatic fibrosis can occur. A recent study has shown that children with biliary atresia suffer a high risk of malnutrition which can negatively affect the course for those who undergo liver transplantation [5]. Children who survive into young adulthood with their native liver need close follow-up because chronic cholangitis or development of portal hypertension can result in a late need for liver transplantation [6].

## Study Questions

1) Question 1: Under the Kasai classification, which anatomic type is a child who has an obliterated common hepatic duct, a patent common bile duct, a patent cystic duct, and a cystic structure at the hepatic hilum.

   a. Type I
   b. Type IIa
   c. Type IIb
   d. Type III

Answer: (b) Type IIa. This child's anatomy is most compatible with Kasai Type IIa, also known as "cystic biliary atresia." This anatomic type is associated with a better than average outcome after Kasai portoenterostomy.

2) Question 2: A newborn develops jaundice and acholic stools. Ultrasound reveals small gallbladder, and on HIDA scan, there is no passage of bile from the liver to the extrahepatic bile ducts. Liver biopsy is performed and shows a paucity of intrahepatic bile ducts and cholestasis. Which is the most likely condition?

   a. Neonatal infectious hepatitis
   b. Biliary atresia
   c. Alagille syndrome
   d. Alpha-1-anti-trypsin deficiency

Answer: (c) Alagille syndrome. There are various conditions that could cause jaundice in a newborn. It is important to differentiate biliary atresia, which will require Kasai portoenterostomy, from other conditions. The ToRCH (Toxoplasmosis, Rubella, Cytomegalovirus, Herpes simplex virus) infections cause jaundice through inflammation of the liver. Alpha-1-anti trypsin (AAT) deficiency is a genetic disorder in which an abnormal AAT protein causes liver damage because it cannot be released from the liver, causing liver injury and cirrhosis. Alagille syndrome is a genetic disorder in which a paucity of interlobar bile ducts causes cholestasis and jaundice. The majority of children with Alagille syndrome have a mutation in the JAG1 gene.

## References

1. Wildhaber BE. Biliary atresia: 50 years after the first Kasai. ISRN Surg. 2012. (Online) https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3523408/pdf/ISRN.SURGERY2012-132089.pdf. Accessed 5 Nov 2021.
2. Chardot C, Carton M, Spire-Bendelac, Le Pommelet C, Golmard JL, Auvert B. Prognosis of biliary atresia in the era of liver transplantation: French national study from 1986–1996. Hepatology. 1999;30(3):606–11.

3. Alagille syndrome. Genetics Home Reference. MedlinePlus. US National Library of Medicine, National Institutes of Health US Department of Health and Human Services. Bethesda, MD, 2021. https://medlineplus.gov/genetics/condition/alagille-syndrome/ . Accessed 9 Nov 2021.
4. Qipeng Z, Fang Y, Yilin Z, Gengxin L, Mengdi L, Xiaoli H, Jianghua Z. The favorable prognosis of cystic biliary atresia may be related to early surgery and mild liver pathological changes. Pediatr Surg Int. 2021., (Online), https://pubmed.ncbi.nlm.nih.gov/34618182/ . Accessed 5 Nov 2021.
5. Boster JM, Feldman AG, Mack CL, Sokol RJ, Sundaram SS Malnutrition in biliary atresia: assessment, management and outcomes. Liver Transpl. 2021. (Online), https://pubmed.ncbi.nlm.nih.gov/34669243/ . Accessed 5 Nov 2021.
6. Namgoong JM, Hwang S, Ahn CS, Moon DB, Ha TY, Song GW, Jung DH, Park GC, Yoon YI, Kim KM, Lee SG. Indications and outcomes of liver transplantation. Korean J Transplant. 2021;35(3):177–82.

# Sacrococcygeal Teratoma

**Danielle Dougherty and Marc S. Lessin**

**Key Points**

1) Sacrococcygeal teratoma is a relatively rare tumor that occurs in infants and children.
2) Teratomas are germ-cell tumors which present with embryonic components containing at least one of the three components of fetal tissue consisting of endoderm, mesoderm, and/or ectoderm.
3) Tumors are classified as external, internal, or mixed; sub-classified as predominantly internal or external.
4) Large sacrococcygeal teratomas may be diagnosed prenatally. High-output cardiac failure related to tumor blood flow may result in fetal hydrops.
5) Most sacrococcygeal teratomas diagnosed in newborns are benign, whereas later diagnosis is associated with malignancy.
6) Alpha fetoprotein is usually elevated but is not useful in the newborn period because of baseline elevation.
7) Treatment requires complete resection including excision of the coccyx.

1. Epidemiology: Sacrococcygeal teratoma is a relatively rare tumor of infancy and childhood. It occurs most commonly in neonates and is largely benign. It is the most common tumor in the newborn. It may be diagnosed in utero. Those that occur in older children or which were not diagnosed because they were intrapelvic are more likely malignant related to the yolk sac component.

   a. Incidence: The occurrence is approximately 1 in 35,000 births. The disease is four times more common in females than males. The incidence of malignancy

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for sacrococcygeal teratoma in the newborn period is about 10% which increases to about 70% by age 4.

b. Etiology: The etiology of teratomas is unknown. A genetic basis of teratomas has not been identified. The hallmark of the disease is presence of at least one germ-cell layer and often more than one germ-cell layer, namely, endoderm, mesoderm, or ectoderm.

2. Pathophysiology:

   a. Anatomy: Sacrococcygeal teratomas are tumors which occur in the presacral area. They may be exophytic, pelvic, intra-abdominal, or a combination of external and internal. They are always in continuity with the coccyx. The blood supply is primarily from the middle sacral artery. The split notochord syndrome described by Currarino and known as Currarino's triad is an autosomal dominant condition consisting of vertebral anomaly involving the sacrum, anorectal anomalies, and presacral mass. The presacral mass is not always a teratoma. Contrary to late diagnosis of sacrococcygeal teratomas beyond the newborn period, the risk of malignancy in a teratoma associated with the Currarino triad is rare.
   b. Classification schemes: (Fig. 1)

      Type I: Predominantly external
      Type II: External with intrapelvic extension
      Type III: External with pelvic and intra-abdominal extension
      Type IV: Entirely internal

3. Clinical features: Large sacrococcygeal teratomas which are external may be diagnosed prenatally. They may have both solid and cystic components. High-output cardiac failure may result in hydrops due to large shunts within the tumor. Tumor rupture or bleeding may occur with large lesions. Urogenital anomalies such as hypospadias and uterine or vaginal duplications have been associated with sacrococcygeal teratoma.
4. Diagnosis:

   a. Symptoms: The primary presentation is a large external sacrococcygeal mass. The skin is often thinned out over the lesion. Pelvic teratomas which present later may present as constipation and dysuria. The differential diagnosis includes myelomeningocele.
   b. Examination: External sacrococcygeal teratomas are obvious (Fig. 2). A rectal examination should be carried out to evaluate the extent of tumor. Radiographic studies are essential to evaluate extent of the tumor.
   c. Laboratory tests: Routine laboratory testing should include complete blood count, alpha fetoprotein, beta human chorionic gonadotropin, and CA-125. Note that alpha fetoprotein levels can normally be in the 100,000 range in normal neonates so these are not diagnostic. Normal levels of alpha fetoprotein are not achieved until 6–8 months of age.

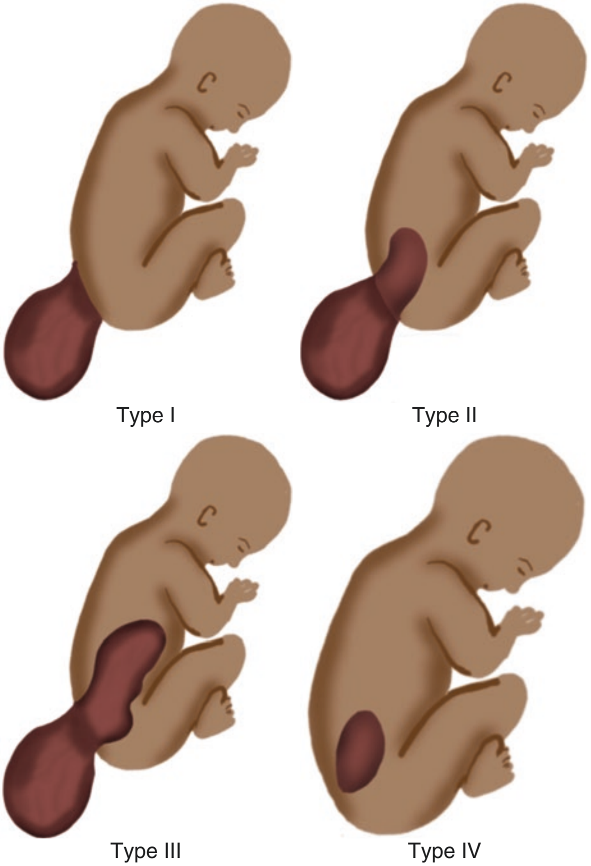

**Fig. 1** There are four types of sacrococcygeal teratomas. Type I is predominantly external. Type II is external with intrapelvic extension. Type III is external with pelvic and intra-abdominal extension. Type IV is entirely internal. (Created by Danielle Dougherty M.D.)

   d. Radiographic studies: Ultrasound, CT, and magnetic resonance imaging have all been used to determine the intrapelvic extent of tumor. Magnetic resonance imaging with arterial and venous phases are useful for determining blood supply.

5. Treatment:

   a. Initial measures: The initial management is observation, cardiac echocardiogram, laboratory evaluation, and imaging to determine extent of pelvic or intra-abdominal involvement for the purpose of preoperative planning. In utero intervention has been performed including the EXIT procedure for large teratomas with impending hydrops. Fetuses with large lesions should not be delivered vaginally to avoid risk of rupture.
   b. Subsequent measures: Excision of mass is not an emergency. The child should be stabilized with intravenous fluids and imaging to determine approach based on extent of the tumor.

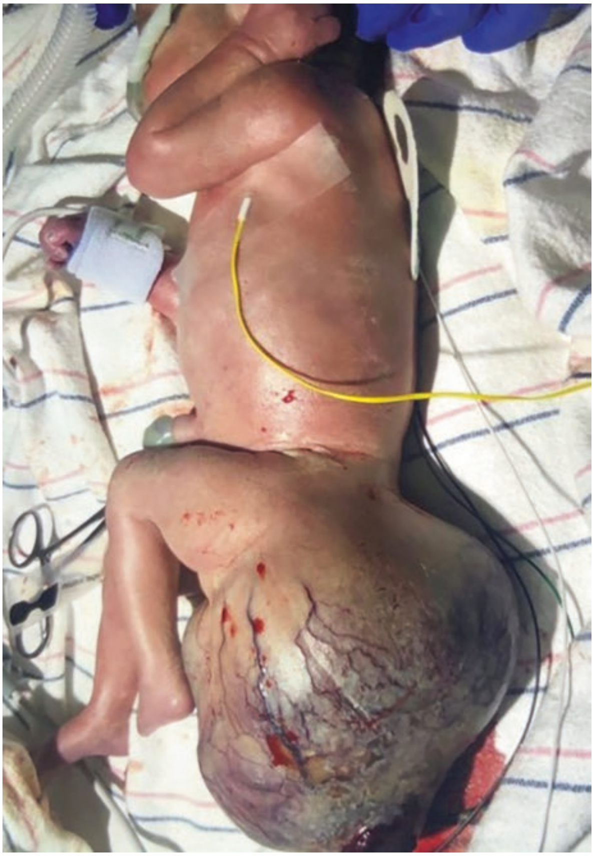

**Fig. 2** Large sacrococcygeal teratoma with partial rupture (Source: Muriel Cleary M.D. personal files)

c. Preoperative preparation: The procedure is done under general anesthesia with endotracheal intubation. Adequate intravenous access is crucial. An arterial line is recommended. Packed red blood cells or whole blood should be available. A gastric decompression tube and urinary catheter should be placed. Depending on extent of pelvic involvement, the procedure may require access to the abdomen. In these situations, the entire body should be circumferentially prepped so that the infant can be turned over after mobilization of the intrapelvic or intra-abdominal tumor or if the approach is posterior so that the infant could be turned over should abdominal access become necessary to control blood supply.
d. Operative treatment: Most sacrococcygeal tumors can be approached from the back with child in the prone position (Fig. 3). If there is a significant pelvic or intra-abdominal component, this part of the tumor is first mobilized by an abdominal approach which may be done laparoscopically. This approach allows for control of the blood flow to the tumor by ligating the middle sacral artery or occasionally one or both internal iliac arteries (Fig. 4). Encircling the aorta has been described to control severe bleeding should it be encountered. The incision used is a Chevron incision resembling an inverted "V" preserving adequate skin for closure as the skin is not normally infiltrated by

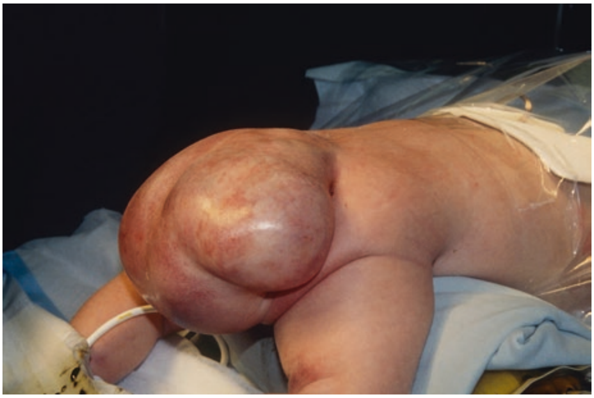

**Fig. 3** Prone positioning for resection of Type 1 sacrococcygeal teratoma (Source: Marc Lessin M.D. personal files)

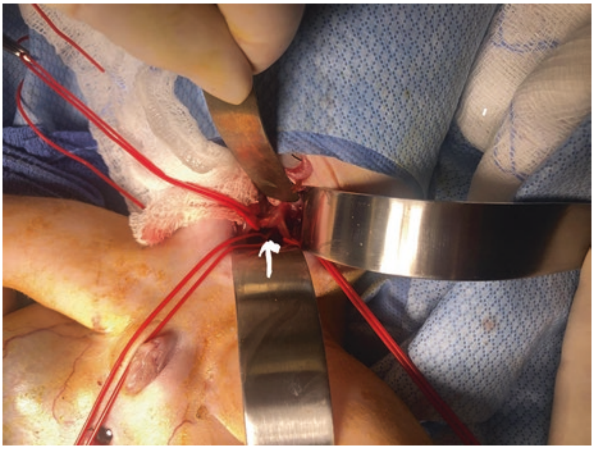

**Fig. 4** Ligation of the middle sacral artery prior to resection. Arrow points to middle sacral artery. Right and left common iliac arteries isolated with vessel loops (Source: Muriel Cleary M.D. personal files)

tumor. Once the tumor is resected, excess skin is removed. The closure should bring the anus posterior from its displaced anterior position and should not be under undue tension (Fig. 5).

e. Postoperative treatment: Postoperative fluids should be continued and the child NPO until bowel function returns. Children should be followed up with serial alpha fetoprotein levels to insure that they return to normal. Normal levels should be achieved in the newborn by 8 months of age. Rectal examination and ultrasound should be performed on a regular basis until at least 3 years of age.

f. Potential complications: Tumor lysis syndrome has occurred with resection. Bleeding as the result of unrecognized arterial supply from the middle sacral artery or internal iliac arteries has been reported prompting sterile preparation of the lower body prior to surgery so that vascular control can be obtained from the abdominal component. Anal transposition can result in problems with stooling due to inadequate motor muscle complex around the anus. Recurrent tumors occur in as many as 20% and tend to be malignant. Most recurrences occur within 3 years of operation.

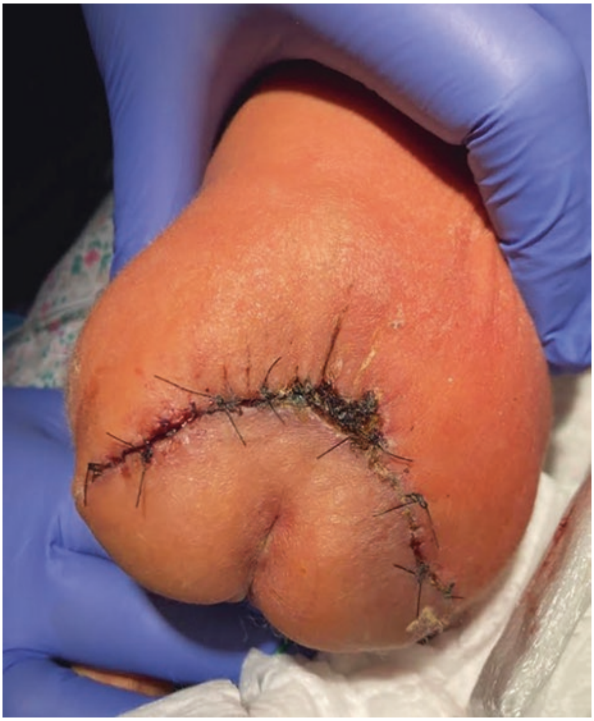

**Fig. 5** Closure of Chevron incision after resection of sacrococcygeal teratoma seen in Fig. 1 (Source: Muriel Cleary M.D. personal files)

## Additional Notes

Teratomas are tumors which derive from germ-cell layers. Traditionally elements of three germ-cell layers were needed to be present, but this is no longer required for diagnosis. They are thought to arise from primordial germ cells. The word is derived from the Greek root meaning “monster” as there may be developed fetal body parts in these tumors. The tumors are primarily midline. Sacrococcygeal teratomas are a specific subcategory of this midline tumor occurring in the sacral–coccygeal region. There are two common scenarios in which sacrococcygeal tumors present. One is in the form of a large and external mass in a neonate, and the second is a relatively hidden mass in the pelvic of a child who typically presents with symptoms of bowel and bladder compression.

Sacrococcygeal teratomas are the most common congenital tumor in neonates and are often detected in utero during routine prenatal screening. They are rarely malignant, but there is a reported mortality rate ranging from 13 to 23%. Mortality is usually caused by either cardiac failure or exsanguinating tumor hemorrhage. This has been associated with tumors that are larger in size, have increased vascularity, and/or demonstrate a rapid growth rate. Tumors that are identified on routine prenatal ultrasounds are typically identified by an echogenic, complex mass seen at the level of the coccyx. Further imaging, such as Doppler ultrasound and MRI, can help delineate feeding vessels and relationship to adjacent structures. It is also important to evaluate for the presence of hydrops, placentomegaly, and cardiac failure, all of which signify a worse prognosis. Management options depend on all of these factors. Fetuses with small sacrococcygeal teratomas that demonstrate none of the risk factors described above may undergo a vaginal delivery and then excision of the tumor within the first few days of life after adequate resuscitation. Fetuses with larger tumors, such as those greater than 5 cm, have been recommended to undergo elective Cesarean delivery.

For fetuses that demonstrate risk factors such as high-output cardiac failure due to a highly vascular tumor, options include elective early delivery and fetal interventions such as tumor resection, laser ablation, drainage of cystic portions, and alcohol sclerosis. Overall, fetal interventions are reserved for select cases in fetuses that are predicted to have a very high mortality rate without intervention. They are performed only at specialized institutions by highly trained surgeons and have a survival rate reported between 30 and 50%.

The second main presentation of sacrococcygeal tumor is a child who has a relatively more obscure tumor in the pelvic region and often presents with symptoms of bladder or rectum compression. Some of these patients have been treated unsuccessfully for voiding issues prior to discovery of the tumor. These tumors are more likely to be malignant but can be successfully treated with complete resection and perioperative platinum-based chemotherapy. Prognosis is good, even in cases of metastasis, and 5-year survival of malignant tumors has been reported to be as high as 89%.

It is important that all patients with sacrococcygeal tumors have close oncologic follow-up. Recurrence can occur and is most common within the first 3 years after the initial operation. Recurrence is associated with incomplete tumor resection at the original operation and standard surgical practice emphasizes complete en bloc resection of the tumor with the coccyx and avoidance of tumor spillage. Long-term follow-up should also be done to evaluate for development of neuropathic bowel or bladder issues, which has been reported in 35–41% of patients after their treatment.

## Study Questions

1) Question 1: All of the following tumor markers are used in the diagnosis of sacrococcygeal teratoma except:
   a. Alpha fetoprotein (AFP)
   b. CA-125
   c. CA-19
   d. Beta human chorionic gonadotropin (BHCG)

Answer to Question 1: C. Alpha fetoprotein and CA-125 but not CA-19 have been used in follow-up of sacrococcygeal teratoma. Beta HCG should be obtained in patients with sacrococcygeal teratoma and may be associated with precocious puberty.

2) Question 2: The patient presents with constipation. Physical examination shows anal stenosis and X-rays demonstrate partial agenesis of the sacrum. All of the following associated anomalies might be suspected except?

   a. Hirschsprung disease
   b. Presacral tumor
   c. Bicornuate uterus
   d. Tethered cord

Answer to Question 2: A. The patient presents with constipation and is found to have anorectal stenosis. X-ray demonstrating partial sacral agenesis should raise suspicion of split notochord syndrome known as Currarino's triad. Currarino's triad consists of anorectal malformation, usually anal stenosis, sacral anomaly, and presacral tumor. Uterine anomalies are also common. Currarino's triad is an inherited condition which is autosomal dominant inherited. The most common presenting symptom is constipation with anorectal malformation. The condition has been misdiagnosed as Hirschsprung disease. The most common tumor is teratoma followed by anterior myelomingocele.

3) Question 3: Which of the following describes an acceptable follow-up plan for a newborn after successful resection of a 3-cm benign sacrococcygeal teratoma?

   a. Annual history and physical.
   b. No surgical follow-up is needed in the case of a small (< 5 cm) and benign tumor.
   c. Imaging (ultrasound or CT) and tumor markers every 6 months for the first 2 years then annually thereafter.
   d. Tumor markers annually for the first 3 years.

Answer to Question 3: C. All tumors have a risk of recurrence and recurrence can be associated with malignancy even if the original tumor removed was benign on pathology. Recurrence is most common within the first 3 years after the original operation and it is important to have adequate follow-up with imaging and tumor markers.

4) Which artery supplies a sacrococcygeal teratoma?

   a. Inferior rectal artery
   b. Middle sacral artery
   c. Internal iliac artery
   d. Internal pudendal artery

Answer: B: The middle sacral artery, which is a direct branch off of the aorta at the level of the iliac bifurcation, provides blood flow to sacrococcygeal tumors and is always divided at surgery.

## Further Reading

Currarino G, Coln D, Votteler T. Triad of anorectal, sacral and presacral anomalies. Am J Roentgenol. 1981;137:395–8.

De Corti F, Sarnacki S, Patte C, Mosseri V, et al. Prognosis of malignant sacrococcygeal germ cell tumours according to their natural history and surgical management. Surg Oncol. 2012;21(2):e31–e7.

Khalil BA, Aziz A, Kapur P, Humphrey G, Morabito A, Bruce J. Long-term outcomes of surgery for malignant sacrococcygeal teratoma: 20-year experience of a regional UK centre. Pediatr Surg Int. 2009;25(3):247–50.

Kitagawa H, Pringle KC. Fetal surgery: a critical review. Pediatr Surg Int. 2017;33(4):421–33.

Kremer ME, Wellens LM, Derikx JP, van Baren R, et al. Hemorrhage is the most common cause of neonatal mortality in patients with sacrococcygeal teratoma. J Pediatr Surg. 2016;51(11):1826–9.

Pauniaho SL, Tatti O, Lahdenne P, Lindahl H, et al. Tumor markers AFP, CA125, and CA 19-9 in long term follow-up of sacrococcygeal tumors in infancy and childhood. Tumor Biol. 2010;31:262–165.

Peiró JL, Sbragia L, Scorletti F, et al. Management of fetal teratomas. Pediatr Surg Int. 2016;32:635–47. https://doi.org/10.1007/s00383-016-3892-3.

Rescorla FJ. Teratomas and other germ cell tumors. In: Pediatric surgery. In: Elsevier; 2012. p. 507–16.

Wu JT, Book L, Sudar K. Serum alpha fetoprotein levels (AFP) in normal infants. Pediatr Res. 1981;15(1):50–2.

# Patent and Persistent Ductus Arteriosus

**Gerhard Ziemer and Cherish Boehm**

**Key Points**

1) The ductus arteriosus is a remnant of fetal circulation bypassing the lungs from the pulmonary artery to the aorta. The ductus arteriosus connects the proximal left pulmonary artery to the descending aorta. Rarely, the ductus is on the right side. Depending on pulmonary vascular resistance (normally declining within days), postnatal blood flow may be right to left or left to right.
2) First-line therapy for patent ductus arteriosus in symptomatic premature neonates is medical with non-steroidal anti-inflammatory drugs (NSAIDs), which as anti-prostaglandins cause ductal closure by constriction. Neonatal cardiac defects with ductal-dependent systemic blood flow (hypoplastic left heart, interrupted aortic arch, etc.) or ductal-dependent pulmonary blood flow (pulmonary atresia or stenosis, etc.) require prostaglandin (PGE-1) infusion to keep the ductus open.
3) Surgical treatment for closure of the patent and persistent ductus arteriosus is done through a left thoracotomy or in preparation of potential future cardiac surgery via median sternotomy. Thoracoscopic clipping has been done, and interventional techniques with occlusion devices are the first-line treatment beyond the neonatal period, with interventional ductus catheter closure in premature babies on the horizon.

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1. Introduction: The ductus arteriosus, the arterial duct, is an essential part of normal fetal circulation. As a physiological shunt in prenatal life, it allows for about 90% of the right ventricular output (60% of combined cardiac output) to bypass the lungs. In the descending aorta, it adds to about 10% of the combined cardiac output coming from the aortic arch through the isthmus. Functional closure of the ductus arteriosus occurs soon after birth, usually between the first and sixth day of life. This is followed by anatomical closure; an involution of the duct tissue that finally becomes the ligamentum arteriosum, a thin fibrotic structure between the aortic isthmus and the proximal left pulmonary artery. While almost always left sided, in rare cases, a ductus can be on the right side, and there are even cases of bilateral ductus arteriosus reported.
2. Epidemiology: The patent ductus arteriosus is associated with prematurity in the newborn. The ductus arteriosus is patent in approximately 90% of premature infants under 28 weeks gestation and in less than 50% in infants 31–33 weeks gestation. Infants weighing less than 1000 g have over an 80% patency.
3. Etiology/pathophysiology: Histologically and functionally, the ductus arteriosus is a unique vascular structure. In prenatal life, histologically the vessel is a large-sized muscular artery with the media almost completely without elasticity. The cells of the tunica muscularis are located circular with unique receptors, which react to postnatal changes (rising oxygen partial pressure, reduction of prostaglandin level in the blood). The muscle cells promote with their contraction the functional closure in 80% of the newborns mostly in the first 24 h postnatally. Additionally, the intima of the ductus has a very well-developed gelatinous matrix, which seals the closing lumen as the muscle contracts. The final closure occurs within weeks caused by apoptosis of the muscular cells and proliferation of fibrotic tissue. At first the closure is completed at the side of the pulmonary artery; an ampulla-like bulging remains on the aortic side for a few months. The early phase of the closing process can be completely reversed (up to 2 weeks of life) by a postnatal infusion of prostaglandin. This may be necessary in patients with certain congenital heart defects with ductal-dependent systemic blood flow (hypoplastic left heart, interrupted aortic arch, critical coarctation of the aorta, etc.) or ductal-dependent pulmonary blood flow (pulmonary atresia or stenosis, etc.) to maintain flow in the ductus arteriosus to either maintain organ and extremity perfusion or oxygenation. On the other hand, the muscular cells in a ductus of a premature baby show a limited ability to react to oxygen, which is one reason for the high rate of a patent ductus in this group of patients.

   Patency of the ductus arteriosus after birth can result in left-to-right shunts exposing the lungs to systemic pressures. This leads to decreased organ perfusion and puts the newborn at increased risk for necrotizing enterocolitis and kidney dysfunction.

   A patent ductus has the potential to spontaneously close for up to 3 months of life. Thereafter, spontaneous closure is unlikely, and the open ductus must be considered persistent. The persistent ductus arteriosus is a distinct pathological–clinical entity.

Patent and persistent ductus arteriosus need to be separately discussed for different age groups and anatomic status with distinct therapeutic approaches: (1) premature babies, (2) infants/children, (3) aneurysmal duct, and (4) infected duct.

4. Anatomy: The ductus arteriosus is a fetal structure which bypasses the lungs by providing an outflow tract for the right ventricle. The ductus arteriosus connects the pulmonary artery bifurcation or the proximal left pulmonary artery to the descending aorta.
5. Diagnosis

   A. Symptoms: Left to right shunting leading to various degrees of cardiorespiratory failure, with cardiomegaly, and eventually pulmonary edema and hypoxia requiring continued mechanical ventilation with high oxygen requirements.
   B. Examination: Continuous murmur over the left precordium known as machinery murmur and widened pulse pressure.
   C. Radiographic studies: The diagnosis of ductus arteriosus is based on 2D echocardiography demonstrating diastolic flow reversal in descending aorta.

6. Patent ductus arteriosus of the premature baby

   A. Indications for closure: The patent ductus arteriosus of prematurity is due to a delayed closure of a histologically normal duct. The ductal tissue is immature, which postnatally makes the ductal myocytes not react physiologically, as it would occur in term babies. Depending on the hemodynamic influence of the patent duct in the individual patient, closure is indicated (increased left heart volume load by a large left/right shunt with persistent pulmonary hypertension). An early sign preceding clinically manifest critical abdominal and/or renal perfusion is the holodiastolic descending aorta flow reversal as seen in echocardiogram studies.
   B. Medical closure: Inhibition of prostaglandin synthesis as a medical therapy was introduced in 1976 with indomethacin, and because of positive results and reduced risks, the medical treatment (now with any type of newer nonsteroidal anti-inflammatory medication) remains the primary therapy in premature neonates. Recent studies have found that ibuprofen is equally effective in closure of the ductus arteriosus and has reduced risk of necrotizing enterocolitis and renal insufficiency. More recently, acetaminophen has been used in an attempt to decrease the incidence of necrotizing enterocolitis associated with indomethacin. Despite its widespread use in premature infants less than 1000 grams, there is a lack of sufficient data as to whether primary surgery or primary medical therapy should be preferred. The only randomized controlled trial of very early prophylactic surgical duct closure is more than 30 years old, leading to a 66% reduction of necrotizing enterocolitis in primary patent ductus arteriosus (PDA) closure. That 1989 study

showed no survival advantage. A meta-analysis by the Cochrane Database in 2013 found there was not enough data to conclude whether surgical ligation or medical treatment with indomethacin is preferred as the initial treatment for symptomatic PDA in preterm infants, primarily relying on a 1983 study!

C. Surgical closure: If possible, the operation should take place in the neonatal intensive care unit (NICU) because of the unstable thermoregulation and the often-critical respiratory conditions in premature babies. The procedure in the NICU maintains the therapeutic environment but also fosters uninterrupted care and monitoring throughout the procedure by an experienced neonatologist and NICU staff.

During the procedure, ECG and pulse oximetry of the upper and lower extremities should be maintained and are often part of routine NICU care. The baby is positioned on the right side. Axillary roll is often placed but is optional. The posterolateral thoracotomy should be kept short due to the extreme elasticity of the tissue and may be no longer than 1.5 inches in premature infants with a weight of <1000 g. A muscle-sparing thoracotomy is preferred, but not always achievable. The intercostal muscle is divided above the rib. In the extrapleural exposure, cotton swabs are used to separate the parietal pleura from the chest wall in a lateral to posteromedial direction. Even though the extrapleural approach is often used, it has no proven advantage over a transpleural approach. The exposure of the site is obtained by careful retraction of the inflated and relatively stiff lung. This maneuver can compromise the cardiac function, and close monitoring regarding the development of bradycardia is required.

In case of a repetitive compromise of the circulation and decrease of the heart rate <100/min, which may occur very rapidly, all instruments should be removed out of the chest immediately, and a few minutes of recovery with the left lung expanded, should be allowed for. The dissection should be minimized to local incisions of the mediastinal pleura cranial and caudal of the ductus adequate to apply a clip. The edematous subpleural tissue of very symptomatic patients further simplifies the dissection.

In many cases, external diameter of ductus arteriosus is as big as descending aorta. Circumferential dissection of the ductus is not only unnecessary but also should be avoided for isolated ductus closure in premature infants. The ductal tissue is extremely fragile so that any direct manipulation should be avoided. The adventitia of the aorta may be grasped and retracted to provide additional length for clip placement. The recurrent laryngeal nerve branch of vagus nerve loops around the ductus ventral and should not be manipulated. The clip should be applied midway of the duct to avoid any narrowing to the aorta or left pulmonary artery (Fig. 1). The clip should not entrap the recurrent laryngeal nerve as this will result in ipsilateral vocal cord paralysis. Pulse oximetry system of the lower extremities is useful to confirm that there is no compromise of the aorta. The clip should completely occlude the ductus. In case of an apparent incomplete closure, the

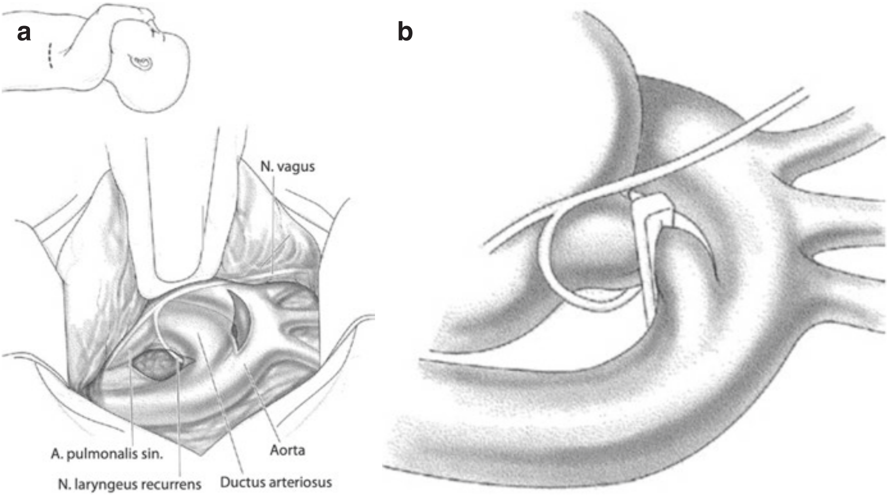

**Fig. 1** (**a**), (**b**) Surgical site for a patent ductus arteriosus in a premature baby. After minimal dissection above and below the ductus, only one clip is applied in the middle of the ductus. (Source: Ziemer G, Haverich A, editors., Operations on the heart and great vessels in adults and children, Springer, 2017).

removal of the clip is not recommended because of the risk of duct laceration. Instead, a second clip must be placed. If the application of the first clip induces any bleeding of the ductus, a second and ideally a third clip applied on each side of the original clip. Prior to application, the clip applier should be tested for undistorted clip application on a string outside the operating field before using to close the duct.

In a standard situation, the chest should be closed without a pleural drain. Preplaced para-costal sutures are tied during a forced inflation of the lungs, in order to guarantee complete expansion of the lungs. Irrigating the wound with normal saline during the first ventilations after chest closure may detect or exclude an airleak. No attempt should be made to close small air leaks. In these cases, a pleural drain should be inserted before closure. The fascia is closed with a running suture, and the skin closed with a running subcuticular with application of Dermabond or Steri-Strips.

D. Interventional closure: Currently, interventional closure efforts in premature infants weighing less than 1000 g is being carried out at several institutions. Theoretical benefits of “avoiding surgery” must be weighed against the requirement of taking patients out of the NICU for procedure. Also, late long-term effects of femoro-iliac vein occlusions, which may be acutely tolerated, are a potential complication (post-thrombotic syndrome).

E. Complications of surgery:

    i. Most frequent complication is injury of recurrent laryngeal nerve (<10%).
    ii. Intraoperative bleeding complications are rare with clip application.

iii. While classic ligation can lead to recanalization in patients beyond prematurity, the mechanical ductus closure (clip or ligation) in premature babies is followed by anatomical ductus closure as the child grows to full post-conceptual maturity.

iv. Chylothorax is a rare complication, and it will appear in premature infants as mere lymph leak, due to lack of enteral feeds in symptomatic patients. It may require secondary clip closure of leak. Octreotide has also been used with success.

v. Inadvertent ligation/closure of left pulmonary artery and the aorta has been reported.

vi. Left-sided diaphragmatic paralysis has been reported due to injury to the phrenic nerve.

7. Persistent ductus arteriosus in infants and children: The persistent ductus arteriosus is a distinct pathological–clinical entity. In contrast to the normal ductal wall tissue, elastic fibers in the media are well developed. It almost resembles what can be seen in the media of the normal aorta. For that reason, persisting patency of the duct cannot be explained by a delayed or incomplete closing, but rather with a different patho-histology.

   A. Indication for closure: This is the typical aorta-like ductus arteriosus which depending on its size and effectiveness can cause a significant left-to-right shunt leading to heart failure with or without increased pulmonary pressure. Beyond the third month of life, the ductus does not show any tendency toward a spontaneous closure, and therefore, the diagnosis of a hemodynamically relevant ductus is considered an indication for transcatheter closure or surgery.

      Doppler sonography can identify a persistent ductus as small as a diameter of 1–2 mm. These tiny vessels are asymptomatic, and prophylactic treatment due to potential complications like endarteritis remains controversial.

   B. Interventional closure: During the last 15 years, interventional closure has been established as a standard procedure in this age group. Open surgery is performed only in exceptional circumstances or due to complications. Since the introduction in the 1970s, interventional techniques for ductus closure have become not only much safer but also more extensive and more expensive. The rate of complications is very low. Acutely embolized devices can be rescued by catheter techniques.

   C. Surgery for embolized device: Surgery to retrieve a lost device out of the pulmonary artery or aorta is far more extensive than a primary simple operative closure or division of the ductus.

      If the dislocation was originally unnoticed and the embolized device may be in its new position for more than 2 weeks, we recommend access via sternotomy and the use of normothermic extracorporeal circulation with the heart beating. A device still sticking in the ductus, however, with a huge residual shunt, may require extracorporeal circulation.

An acutely embolized device usually ends up in the lower lobe arterjy Conditions requiring patency of the ductus arteriosus of the left pulmonary artery. This can be removed directly out of the interlobar fissure (upper/lower lobe) via a posterolateral thoracotomy in the fifth intercostal space and an oblique incision of the artery during a temporary tourniquet closure. In these cases, one needs to also close the ductus.

D. Video-assisted thoracoscopic ductus closure: Video-assisted thoracoscopic ductus closure was introduced in 1993. It has been successful in several series. It offers advantages for postoperative pain and cosmesis (small incisions). The thoracoscopic approach has become less favorable with alternative interventional techniques.

E. Open surgical closure: The classical surgical closure of the ductus is performed through a left posterolateral thoracotomy in the third or fourth intercostal space. The following rule of thumb approximately applies: the lighter the patient, the higher the intercostal space to be selected (<3 kg, third intercostal space; >4 kg, fourth intercostal space; in between gray zone). The intercostal muscle is divided above the rib. The lung is retracted medially. The mediastinal pleura is opened longitudinally over the proximal descending aorta. The ventral edge of the pleura is elevated by stay sutures protecting the lung with a wet gauze pad replacing the lung retractor.

The identification of the ductus may not always be achieved initially. The aortic arch may be mistaken for the ductus. The recurrent laryngeal nerve branching from the phrenic nerve and embracing the ductus from ventral/caudal to dorsal may serve as a guideline. The aorta proximal and distal to the ductus should be dissected circumferentially before the ductus itself is manipulated. Even though it is possible to place two special clamps (Potts clamps) at the ductus in individual cases, if required, it is more convenient to cross-clamp the aorta proximal and distal to the ductus, as it also enables the closure of extremely short and wide structures (Fig. 2).

After transection, both stumps should be sewed by/with double-row polypropylene sutures. In small children, it is only necessary to apply two circumferential sutures, one each on the pulmonary and aortic sides, respectively, thereby avoiding aortic cross-clamping. The descending aorta, however, proximally and distally to the ductus insertion, is prepared for potential cross-clamping if necessary. The ductus is transected in between the tied sutures in small steps. In case of significant bleeding, the cross-clamping has to take place as described above. Often an additional clip application is sufficient, which is performed prophylactically in most cases. An additional suture is rarely needed.

F. Complications of surgery:

   i. Injury of recurrent laryngeal nerve is less frequent than in premature infants. Intraoperative bleeding complications are manageable with proper preparation.

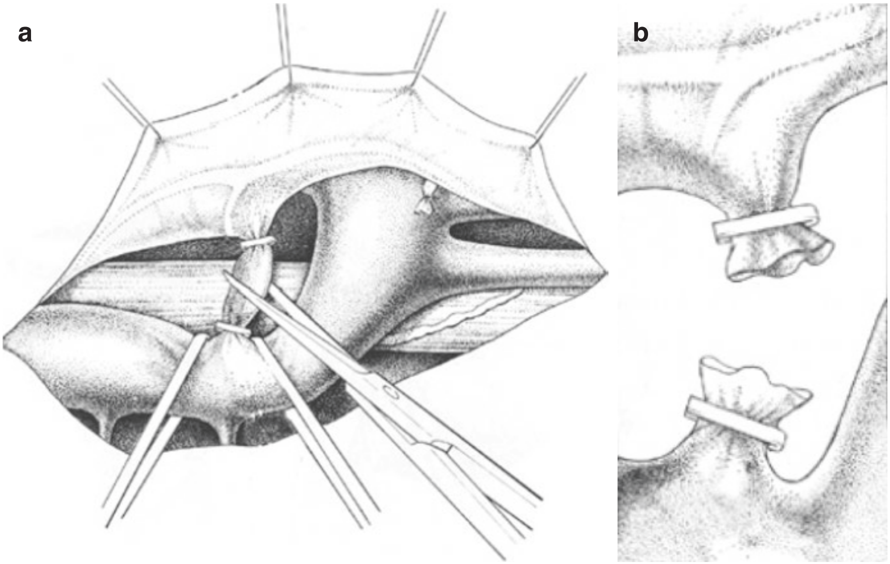

**Fig. 2** (**a**), (**b**) Transection of a persistent ductus arteriosus in infants and children performing purse-string suture closures and if applicable additional clip placements on both the pulmonary and aortic side. (Source: Ziemer G, Haverich A, editors, Operations on the heart and great vessels in adults and children, Springer, 2017)

ii. Classic ligation can lead to recanalization, which is avoided by primary double ligation and some distance between ligatures (division of ductus is preferred technique).
iii. Chylothorax is a rare complication. First-line treatment is medical trial with low fat formula containing medium chain triglycerides. If not successful, secondary clip closure of leak under continuous feeds (preop feeding tube with intubation overnight) for visibility is the second-line approach. Octreotide has also been successful in some cases is particularly useful if there is a contraindication for direct redo surgery.
iv. Inadvertent ligation/closure of left pulmonary artery is reported, as well as left-sided diaphragmatic paralysis due to injury to the phrenic nerve.

8. Conditions requiring patency of the ductus arteriosus for survival: There are three principle hemodynamic situations which require maintenance of post-partum ductal patency in congenital heart disease:

   A) Obstructive right heart lesions, relying on ductus for adequate pulmonary blood flow: severe tetralogy of Fallot-type lesions (subpulmonary stenosis or pulmonary atresia/stenosis with ventricular septal defect).
   B) Obstructive left heart lesions that rely on the ductus for adequate complete or partial systemic blood flow:

    i) Complete: Hypoplastic left heart syndrome (aortic atresia/stenosis, hypoplastic left ventricle). Critical aortic stenosis (high-grade valvar hypoplasia/stenosis with two normal-sized ventricle)
    ii) Partial: Interrupted aortic arch, critical coarctation of the aorta (interrupted aortic arch ductal patency leads to antegrade blood flow in descending aorta by right-to-left shunt from pulmonary artery to descending aorta. In coarctation, the open ductus/relaxed ductal tissue provides unobstructed antegrade blood flow in descending aorta, leading to left-to-right shunt. In this condition, the therapeutic feature is relaxation of intra-aortic ductal tissue in descending aorta).

  C) Cyanotic lesions with inadequate mixing of oxygenated and desaturated blood: The ductus provides exchange of desaturated aortic blood with saturated pulmonary artery blood (d-transposition of the great arteries; d-malposition of great arteries in double outlet right ventricle/subpulmonic ventricular septal effect).

9. Infection of the ductus arteriosus

   Endarteritis inside the ductus and the pulmonary artery is rare. The risk of duct infection, even in the pre-antibiotic era, was only about 1%/year and in the Western world is lower if not nearly nonexistent with current era antibiotics. The objective of surgical therapy would not only be closure of the ductus but also the complete resection of the infected (vascular) parts of the vessel including vegetations. Cardiopulmonary bypass is recommended for this operation.

10. Aneurysm of the neonatal ductus arteriosus

    An aneurysm of the ductus arteriosus may be present prenatally and demonstrable in fetal echocardiography. It also can manifest itself in adults as a late complication of a persistent or incomplete closed ductus, after surgery, intervention, and/or infection.

    The incidence in children with genetic anomalies (trisomy 21, trisomy 13, Ehlers Danlos Syndrome, Marfan's Disease) is higher. Nowadays a primary conservative treatment of watchful waiting is recommended. Regression should be documented by ultrasound performed two to three times daily in the neonate.

    If regression does not occur or a partial thrombosis without closure of the ductus persists (in the area of the aneurysm), surgery should be considered. The principal operative steps are the same as described above, as the aneurysm is constrained within the ductus, and not involving the connecting great arteries. The generally elongated of the ductus is even easier to transect. If thrombus extending into the aorta is documented, the clamping of the isthmus and proximal descending aorta with inspection of the aortic lumen is obligatory.

    Aortic thromboembolism may occur even in premature infants after clip application of a large ductus/ductus aneurysm. Aortic occlusion may be treated by a short-term medically induced rise in the blood pressure, pushing the acute/fresh thrombus peripherally in the bloodstream, ideally to the iliac/femoral vessels where it may dissolve spontaneously.

## Additional Notes

Surgery for a persistent arterial duct was the first successful operation to repair congenital heart disease. After previous work in the laboratory and autopsy room, it was Robert Gross from Boston Children's Hospital, who as a chief resident performed ligation of a symptomatic large-caliber arterial duct via a left-sided thoracotomy in a 7-year-old girl on August 26, 1938. As his chief, Dr. William Ladd, was out of town for summer vacation, he had to ask permission from acting chief, Thomas Langman. Being asked by Dr. Ladd how things were in the hospital, when meeting him shortly thereafter at a sporting event, Gross answered that nothing unusual had happened. When Ladd returned and found out, he fired Gross. This was short-lived as Gross returned shortly thereafter at Ladd's request.

When Porstmann (Charite, Berlin, Germany) closed a persistent arterial duct with catheter technique in an adult in 1966, it became the first congenital heart defect to successfully undergo definitive interventional treatment.

Despite not being accurate, most ductus closures are referred to as "ligations", as the historically first successful ductus closure had been a ligation in 1938. This term should only be used if actual ligations, either done only in premature babies or in infants beyond 3 months and young children, were done. The vast majority of surgical ductus procedures already in Dr. Gross's own practice 1938–1972 (1610 ductus closures) were ductus divisions and nowadays are clip closures in premature babies. A better neutral term would be "ductus closure" as a functional result of any type of surgery.

## Study Questions

1) The ductus arteriosus is a structure in fetal circulation which connects directly:

   a. The aorta to the right ventricle
   b. The vena cava to the aorta
   c. The pulmonary artery to the aorta
   d. The right ventricle to the aorta

Answer to Question 1: (c.) The pulmonary artery to the aorta. The ductus arteriosus is an arterial structure which in normal fetal circulation shunts blood from the right ventricle to the descending aorta. It connects the proximal left pulmonary artery to the aorta. In fetal life, the lungs are not ventilated. Hence, there is no role to perfuse the lungs since they are not the source of oxygen. Oxygenated blood is delivered to the fetus through the placenta. After birth, with the placenta being disconnected, the neonate's first breath leads to a dramatic shift in oxygenation. The ductus is no longer physiologically necessary and usually closes spontaneously within days.

2) Which of the following structures is most frequently at risk during surgery for ductal closure:

   a. The recurrent laryngeal nerve
   b. The left pulmonary artery
   c. The descending aorta
   d. Thoracic duct
   e. All of the above are at same risk.

Answer to Question 2: (a.) The recurrent laryngeal nerve. The ductus arteriosus runs from the left pulmonary artery or sometimes the pulmonary artery bifurcation to the descending aorta. The recurrent laryngeal nerve, a branch of the vagus nerve, loops around the ductus arteriosus. There is a short distance of the ductus between the pulmonary artery and aorta for ductal closure. Injury to the recurrent nerve is the most common structure injured during ductus closure. Injury to the recurrent nerve results in paralysis of the left vocal cord. The left pulmonary artery and aorta have both been inadvertently clipped. The large ductus may be as large as these structures. Identification of the anatomy is crucial to avoid a devastating injury to these arteries. Other injuries include the thoracic duct resulting in chylothorax, injury to the phrenic nerve (retraction injury) with paralysis of the hemidiaphragm, and injuries to the lung.

3) Although clip closure is the preferred surgical procedure for patent ductus arteriosus in premature babies, single ligature may lead to permanent closure also. Single ligature closure, however, is not recommended in closure of persistent ductus arteriosus (beyond three months of age) because:

   a. Tissue in persistent ductus arteriosus is too friable/delicate for manipulation required for ligature.
   b. Closure of persistent ductus arteriosus can never be achieved with single ligature, only.
   c. Single ligature closure of any large vessel has high risk of recanalization.
   d. Ductal ligature always has higher risk for recurrent laryngeal or phrenic nerve damage.

Answer to Question 3: (c.) Single ligature closure of any large vessel has high risk of recanalization. While the patent ductus arteriosus of both the premature and term neonate in the vast majority of cases has normal ductal tissue, capable of spontaneous closure at some point, the persistent ductus arteriosus is a distinct pathological–clinical entity without this capability. In contrast to the normal ductal wall tissue, elastic fibers in the media are well developed. It almost resembles what can be seen in the media of the normal aorta.

Surgical closure in patent ductus arteriosus serves as a bridge until anatomical closure occurs. Clip closure in premature babies has lower bleeding complications than closure with ligature. Single ligature closure of persistent duct has the acute desired effect, but even more than single ligature closure of any artery, is at high risk for recanalization with pulsatile blood flow reaching the ligature site from both sides. Rarification of squeezed arterial wall tissue within the ligature will lead to recanalization over time. Only primary ductus division (preferred) or double ligature with at least short distance between the two ligatures will lead to reliable surgical result in closure of persistent ductus arteriosus.

## Further Reading

Cassady G, Crouse DT, Kirklin JW, Strange MJ, et al. A randomized, controlled trial of very early prophylactic ligation of the ductus arteriosus in babies who weighed 1000 g or less at birth. N Engl J Med. 1989;320:1511–6.

Malviya NM, Ohlsson A, Shah SS. Surgical versus medical treatment with cyclooxygenase inhibitors for symptomatic patent ductus arteriosus in preterm infants. Cochrane Database Syst Rev. 2013;3:CD003951.

Mitra S, Florez ID, Tamayo M, Mbuagbaw L, et al. Association of placebo, indomethacin, ibuprofen, and acetaminophen with closure of hemodynamically significant patent ductus arteriosus in preterm infants: a systematic review and meta-analysis. JAMA. 2018;319(12):1221–38.

Ohlsson A, Shah PS. Paracetamol (acetaminophen) for patent ductus arteriosus in preterm or low birth weight infants. Cochrane Database Syst Rev. 2018:CD10061.

Porstmann W, Wierny L, Warnke H, Gerstberger G, Romaniuk PA. Catheter closure of patent ductus arteriosus. 62 cases treated without thoracotomy. Radiology Clinics of North America. 1971;9(2):203–18.

Stankowski T, Aboul Hassan SS, Seif-Zainab F, Fritsche D, et al. Descriptive review of patent ductus arteriosus by video-assisted thoracoscopy in pediatric population: 7-year experience. J Thorac Dis. 2019;11(6):2555–63.

Zahn EM, Peck D, Phillips A, Nevin P, et al. Transcatheter closure of patent ductus arteriosus in extremely premature newborns: early results and midterm follow-up. JACC Cardiovasc Interv. 2016;9(23):2429–37.

Ziemer G, Kaulitz R. Congenital anomalies of the great vessels, 21.4.1 Ductus Arteriosus Botalli. In: Ziemer G, Haverich A, editors. Cardiac surgery. Operations on the heart and great vessels in adults and children. first Engl ed. Berlin, Heidelberg: Springer; 2017. p. 699–707.

# Neonatal Hyperinsulinemia

**Kody L. Moore and Christopher P. Coppola**

**Key Points**

1) Neonatal hyperinsulinism is a rare congenital disease identified by inappropriate secretion of insulin, resulting in persistent hypoglycemia that can lead to irreversible neurological damage if left untreated.
2) The differentiation of diffuse versus focal subtypes of congenital hyperinsulinism (CHI) is clinically important early on as classification has a significant impact on therapeutic approach to the disease.
3) Fluoro-L-3-4-dihydroxyphenylalanine positron emission tomography (18-fluoroDOPA PET/CT) is considered the gold-standard imaging study which aids in the diagnosis of CHI, and it can also assist with the differentiation of both diffuse and focal classifications.
4) Medical management of neonatal hyperinsulinism can be attempted with diazoxide, a medication that works by keeping the ATP potassium channel open.
5) Approach to CHI varies significantly based on the subtype and severity of the disease, ranging from non-operative medical management to selective partial pancreatectomy to subtotal pancreatectomy.

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1. Background:
    a. Congenital hyperinsulinism (CHI) is a disease process that is characterized by an inappropriate excessive secretion of insulin resulting in persistent, and at times, severe hypoglycemia. There have been several genetic mutations that have been identified as contributing to the cause of CHI; however, many mutations have yet to be fully understood. If left untreated, it can result in severe neurological damage. CHI can be further broken down into both diffuse and focal forms. Rapid diagnosis of neonatal hyperinsulinism is needed to establish appropriate therapy which can include both medical management and at times a surgical approach.
2. Pathophysiology:
    a. Epidemiology
        i. Incidence: Congenital hyperinsulinemia (CHI) has an incidence rate of 1 in 50,000 births. The incidence rate can be as high as 1 in 2500 births in populations of high inbreeding. Most cases are identified within the first few days to weeks of life. However, some cases have delayed presentation at upwards of 6 months of age. Diffuse subtype of the disease is more common (60–70%) as opposed to the focal subtype which is less common (30–40%).
    b. Classification schemes:
        i. Focal: Involves a distinct area within the pancreas consisting of abnormally hyperplastic islet cells with a large amount of B cells which are persistently hyperactive in production of insulin. The lesion is typically described to have irregular borders which are surrounded by normal appearing islets. The focal area typically has very little insulin present due to constant excretion occurring.
        ii. Diffuse: Involves the entire pancreas where there is presence of large B cells that result in persistent hyperproduction of insulin secretion.
        iii. Transient and alternate forms: These forms have various factors including external and syndromic influences.
            1. Maternal diabetes can cause hyperinsulinemic hypoglycemia in the neonatal period.
            2. Those children born with Beckwith–Wiedemann syndrome have a high incidence of hyperinsulinism presentation at birth, often resolving without surgical intervention.
            3. Both Turner and Kabuki syndromes are also linked to hyperinsulinism.
            4. There are several other rare genetic mutations that can lead to neonatal hyperinsulinism, which do not correspond with sensitivity of the ATP potassium channel.

   c. Etiology: There are several known mutations within genes that result in the occurrence of both focal and diffuse forms of congenital hyperinsulinism. The most common mutations result in a loss of function involving ATP potassium channels which results in the inability of adenosine diphosphate (ADP) to affect the pancreatic B-cell. This leads to a continuous stimulation of the cells causing an over secretion of insulin.
   d. Anatomy: CHI involves the insulin-producing cells of the pancreas. Typically, in the form of either a diffuse involvement of all B cells or a focal area of islet cell hyperplasia. Focal lesions can encompass both superficial and deep areas and can be present in all aspects of the pancreas, including head, body, and tail. These lesions are often small, encompassing less than 1 cm in diameter.

3. Clinical presentation:

   a. The main presenting symptom identified in CHI is persistent hypoglycemia with a hallmark of inappropriate insulin ratios.
   b. A wide range of symptoms may also be present in the infant. Both diffuse and focal forms have nearly identical presentations. The symptoms can range from mild to severe, such as subtle lethargy and irritability to severe neurological symptoms of seizures, apnea, and even coma. Earlier onset of symptoms typically corresponds with more severe symptoms. Macrosomia is often present in the neonate.

4. Diagnosis: Laboratory testing will often be the initial indicator of hyperinsulinism. Genetic testing will aid in the confirmation as well as help identify focal versus diffuse disease. Imaging is also valuable with aiding in this distinction. The identification of the diffuse and focal subtypes of CHI is unable to be determined with standard transabdominal ultrasound, CT, or MRI imaging. Invasive interventional sampling has been performed since the 1980s. However, more recently, DOPA PET/CT has been the most precise noninvasive imaging in classifying CHI as well as localizing focal lesions (Fig. 1).

   a. Laboratory tests:

      i. Blood tests showing hypoglycemia of <50 mg/dl will be the initial identifying factor. When present at birth, this will be persistent and lasting >48 h.
      ii. During the initial evaluation of the hypoglycemia, inappropriately elevated insulin may also be seen.
      iii. The patient will proceed with a glucagon stimulation test. An inappropriate response, a rise of glucose >30 mg/dl, is consistent with neonatal hyperinsulinism.
      iv. Once an initial diagnosis is given, further genetic testing is needed to aid with both prognostic factors, subtype identification, and anticipation of clinical course of treatment.

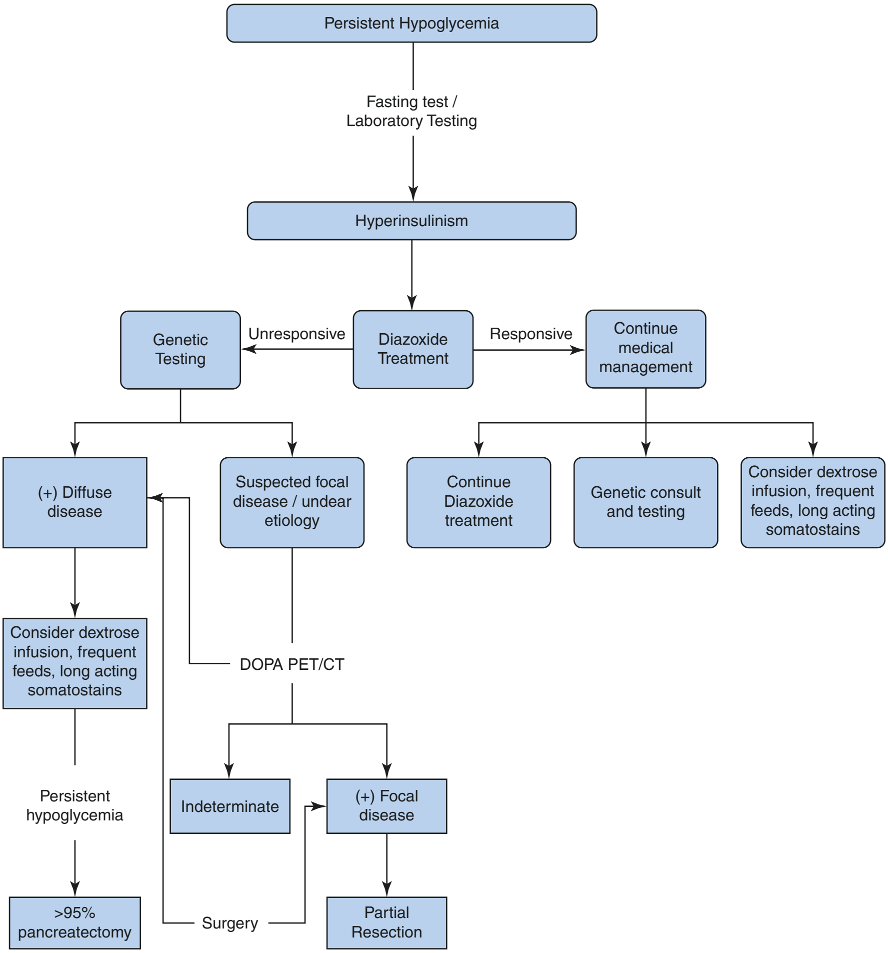
Persistent Hypoglycemia
Fasting test / Laboratory Testing
Hyperinsulinism
Genetic Testing
Unresponsive
Diazoxide Treatment
Responsive
Continue medical management
(+) Diffuse disease
Suspected focal disease / undear etiology
Continue Diazoxide treatment
Genetic consult and testing
Consider dextrose infusion, frequent feeds, long acting somatostains
Consider dextrose infusion, frequent feeds, long acting somatostains
DOPA PET/CT
Persistent hypoglycemia
Indeterminate
(+) Focal disease
>95% pancreatectomy
Surgery
Partial Resection

**Fig. 1** Treatment algorithm for congenital hyperinsulinism (adapted from Adzick and Laje [3])

b. Radiographic studies:

   i. 18-fluoroDOPA PET/CT: considered the gold-standard imaging for evaluation of CHI. Used for differentiation of both diffuse and focal subtypes and is considered the most precise imaging available.
   ii. Venous sampling: This method was initially used prior to development of the PET/CT scan to help localize focal lesions. Due to the invasive approach and inability to guide surgical approaches, it is often no longer recommended in diagnostic evaluation.
   iii. Intraoperative ultrasound: This study can guide in finding focal lesions during surgical cases as well as identify the path of the pancreatic duct during resections, aiding in decreasing complications of ductal injury.

5. Initial management: With any newborn found to have persistent hypoglycemia, it is important to stabilize the infant to normoglycemia to resolve associated symptoms as well as limit potential for neurological damage. A goal of maintaining blood glucose levels >70 mg/dL is recommended.
6. Treatment of neonatal hyperinsulinism:

    a. Medical management: Once hyperinsulinism has been diagnosed and hypoglycemia has been controlled, the medication diazoxide is typically initiated. This medication can manipulate the ATP potassium channel in a way that allows it to remain open, thus preventing persistent excretion of insulin in a large portion of cases. Once a patient shows response to the medication, a trial of fasting is typically performed. If severe hypoglycemia does not resolve, the patient is maintained on the medication with periodic dosing reassessments to see if a decrease or even a total discontinuation of the medication can be performed. When a patient is resistant to diazoxide and does not show a response that results in resolution of the hyperinsulinism, further evaluation is warranted to determine candidacy for surgical treatment. In this situation, there is often a course of frequent feedings and at times glucose infusions to control the hypoglycemia. Trials of long-acting somatostatins can also be considered as they can inhibit the production of insulin as well.
    b. Operative treatment: In those infants who are refractory to medical management, or have an identifiable focal disease, partial to subtotal pancreatectomy is to be considered. Determining diffuse versus focal CHI is crucial to the surgical approach. Often when a focal lesion is identified, surgical excision of this lesion results in a cure of the hyperinsulinism. For those with diffuse disease, who have failed medical management, decreasing the amount of insulin that is secreted is the goal of surgery. This often requires a near-total pancreatectomy. It is necessary to have both a surgeon with experience in pancreatic surgery and a pathologist comfortable with intraoperative diagnosis of focal lesions.
    c. Operative approach: A transverse supraumbilical laparotomy is typically performed. Laparoscopic approach can be considered in instances where a focal distal pancreas lesion has been identified on pre-surgical imaging. If there is known focal disease identified on pre-surgical imaging, then this area of the pancreas is exposed during a laparotomy approach. Often full exposure of the pancreas is needed. If focal disease is present, these areas can typically be identified with palpation and visualization of the lesion(s). It is advised that biopsies are taken from the head, body, and tail of the pancreas and sent for frozen pathology to confirm or rule out diffuse pathology.

        i. Diffuse disease confirmed intraoperatively with frozen section biopsies will require a near-total pancreatectomy being performed, leaving a small portion of pancreas between the common bile duct and duodenum (upwards of 98% of pancreas removed). Preservation of splenic vessels and the common bile duct (CBD) is necessary. It is highly recommended that these patients also receive a gastrostomy tube placement at the time of the operation to aid in postoperative care.

    ii. When initial biopsies show a histologically normal pancreas, the search for focal lesion(s) should begin. Pre- and intraoperative imaging greatly help with finding these lesions. Once a focal lesion is identified, the determination of superficial versus deep lesion is warranted. Simple resections can be performed for superficial lesions. For deep lesions, a partial pancreatectomy is performed. This is determined by the exact location of the lesion. If a deep lesion is present in the head of pancreas, a Roux-en-Y procedure may be warranted. The use of frozen biopsies aid in the ability to have surgically clear margins prior to the end of the case.

        1. Consider the usage of intraoperative sonography to further aid in the identification of focal lesions. Mobilization of the pancreas during the operation can distort the location of a focal lesion seen preoperatively on initial PET/CT imaging, making it difficult at times to identify the location of a previously known lesion. Furthermore, the use of high-resolution sonography can readily identify both the pancreatic duct and surrounding blood vessels, allowing for a reduction in the risk of injuring these important structures. Lastly, intraoperative ultrasound has shown the ability to visualize miniscule extending structures of a focal lesion not readily seen on preoperative PET/CT images. This in turn further aids in providing real-time clear margins without solely relying on frozen biopsies, thus increasing the probability of having a complete resection.

    iii. For atypical and syndromic associated hyperinsulinemia cases that proceed to have a surgical resection, a multidisciplinary treatment approach will be needed on a case-by-case basis.

d. Postoperative treatment: Due to the stress of surgery, reestablishment of glucose infusion is often necessary. Glucose levels need to be monitored at regular intervals in the initial postoperative setting. Once bowel function returns and enteral feeds are resumed, weaning of glucose infusions can be initiated. Once regular enteral feeds have been established and the patient is in a state of euglycemia, a fasting test can be performed to determine if hypoglycemia returns. If a patient cannot be weaned entirely from intravenous glucose, further assessment is needed to determine if additional surgical intervention is necessary.

    i. In certain cases of diffuse CHI, additional medical management is required to control the hypoglycemia, including combinations of frequent feedings, dextrose infusions, and long-acting somatostatins.
    ii. For patients with hyperglycemia and insulin insufficiency, initiation of insulin treatment is often required.

7. Outcomes:
    a. Complications:
        i. One of the main long-term complications that can arise after resection is persistent hypoglycemia. In these patients, further medical management or more aggressive pancreatic resections will be needed.
        ii. Both ductal and vessel injury can occur during a pancreatectomy, most notable are the common bile duct, pancreatic duct, and splenic vessels. The complication of a splenectomy is possible when there is significant injury to splenic vessels.
        iii. Duodenal injury has been reported with diffuse disease and focal lesions involving the head of the pancreas.
        iv. A portion of patients will go on to develop diabetes mellitus often directly corresponding with the amount of pancreas removed. These patients often will require insulin management.

## Additional Notes

The presence of hypoglycemia in the neonatal period is not an uncommon finding. In fact, many causes of hypoglycemia are transient and resolve without the need for surgical intervention. Congenital hyperinsulinism was first coined greater than 50 years ago as "nesidioblastosis." This essentially described what was thought to be an over proliferation of islet cells in the pancreatic duct that lead to the condition which is now known as persistent hyperinsulinemic hypoglycemia of infancy (PHHI). Thanks to the great advances in genetic understanding and testing in recent decades, we now know of at least a dozen different mutations and syndromes that can lead to the neonatal hyperinsulinism. The ongoing advances in the ability to genetically test these infants have also led to an increased ability to separate diffuse cases from focal ones. For the latter, this ability has led to a decreased prolongation of medical therapy in favor of sooner surgical resection, allowing for faster recovery and a cure for many patients.

However, diffuse forms of congenital hyperinsulinism have several different pathways regarding therapy and the decision to proceed with a surgical approach can be difficult. Often the patient and family are dealing with the decision of extensive medical therapy versus an extensive surgery, either of which can have a major impact on the future well-being of the child. Unlike the focal form, the outcomes of surgery during a diffuse case can become unpredictable and differ on a case-by-case basis. Furthermore, a significant amount of the pancreas is often removed during these cases and the goal should be to remove as little of the pancreas as possible to improve the quality of life for a child. In most cases, a cure is not achieved with resection alone, and further medical management is necessary. Initially, it may be worth a trial of a more conservative resection as opposed to that of an extensive one. If medical management afterwards is effective in controlling the hypoglycemia, there may be less risk for the patient to develop diabetes mellitus later in life. If this initial conservative resection fails, additional resection(s) can always be performed.

Advances in minimally invasive surgery have occurred over the last several decades. There is increased use of laparoscopic pancreatectomy for congenital hyperinsulinism, especially with focal disease. Lesions identified in the distal portion of the pancreas are most amenable to this approach. There needs to be caution when dealing with diffuse forms and focal lesions that may be close to the head and body of the pancreas, as there is potential for increased injury to surrounding structures, especially the duodenum and common bile duct. Another potential disadvantage is the loss of tactile ability to feel a focal lesion that may not be visible. Furthermore, the pancreatic duct becomes more at risk the closer the dissection is to the central parts of the pancreas. With advancements in intraoperative ultrasound aiding in identifying this duct, there is lack of ability to use this imaging with laparoscopic approach.

## Study Questions

1) What is the main identifying finding in an infant with congenital hyperinsulinism?

   a. Macrosomia
   b. Transient hypoglycemia
   c. Epilepsy
   d. Persistent hypoglycemia

   Answer:

(d.) Persistent hypoglycemia. Transient hypoglycemia in the neonatal period is not an uncommon finding and typically resolves with the first few days of life. Any infant who has persistent hypoglycemia, especially those with hypoglycemia levels <50 mg/dl, need further evaluation for possible congenital hyperinsulinism as this is a hallmark finding. Both macrosomia and seizures can be seen in an infant with neonatal hyperinsulinemia; however, neither may be present in an infant with congenital hyperinsulinemia.

2) Once a diagnosis of congenital hyperinsulinism is confirmed, the following imaging can be performed to help aid in further evaluation and classification:

   a. Abdominal/pelvic CT with PO and IV contrast
   b. DOPA PET/CT
   c. MRI of abdomen with IV contrast
   d. Complete abdominal ultrasound

Answer:

(b.) DOPA PET/CT. The imaging that is considered the gold standard for evaluation and assistance with classification of congenital hyperinsulinism is 18-fluoroDOPA PET/CT. The PET/CT is the only noninvasive imaging available to help differentiate between diffuse disease and focal forms of hyperinsulinism. With the ability to identify presence of focal lesions, this imaging can help limit the amount of pancreas that needs to be resected, at times helping to avoid subtotal pancreatectomy early on in life. CT, MRI, and ultrasound do not aid in the evaluation and classification of congenital hyperinsulinism.

3) Once hypoglycemia has been controlled, what medication is typically initiated in the initial treatment and evaluation of congenital hyperinsulinism?

   a. Octreotide
   b. Insulin
   c. Diazoxide
   d. Glucagon

Answer:

(c.) Diazoxide. Once normoglycemia is reached, the patient will undergo a diazoxide trial to evaluate for responsiveness. Patients who have an appropriate response to the medication will continue to be managed medically. Those who are unresponsive need further evaluation and management. Octreotide can be useful in patients who are not responsive to diazoxide as it is a long-acting somatostatin and can help inhibit insulin secretion. Glucagon can help to stabilize a patient's hypoglycemia initially and can be used in medical management of those that are not responsive to diazoxide. Insulin is at times needed for patients who go on to develop insulin insufficiency after pancreatectomy, but it is not of use during the initial management of congenital hyperinsulinism.

## Bibliography

1. Adzick NS, De Leon DD, States LJ, et al. Surgical treatment of congenital hyperinsulinism: Results from 500 pancreatectomies in neonates and children. J Pediatr Surg. 2019;54(1):27–32. https://doi.org/10.1016/j.jpedsurg.2018.10.030.
2. Vajravelu ME, De León DD. Genetic characteristics of patients with congenital hyperinsulinism. Curr Opin Pediatr. 2018;30(4):568–75. https://doi.org/10.1097/MOP.0000000000000645.
3. Adzick NS, Laje P. Pancreatic neoplasms in children. In: Carachi R, Grosfeld JL, editors. The surgery of childhood tumors. Springer; 2016. p. 312–21. https://doi.org/10.1007/978-3-662-48590-3_17.
4. Palladino AA, Stanley CA. A specialized team approach to diagnosis and medical versus surgical treatment of infants with congenital hyperinsulinism. Semin Pediatr Surg. 2011;20(1):32–7. https://doi.org/10.1054/j.sempedsurg.2010.10.008.
5. Rahier J, Guiot Y, Sempoux C. Morphologic analysis of focal and diffuse forms of congenital hyperinsulinism. Semin Pediatr Surg. 2011;20(1):3–12. https://doi.org/10.1053/j.sempedsurg/2010.10.010.

6. Pierro A, Nah SA. Surgical management of congenital hyperinsulinism of infancy. Semin Pediatr Surg. 2011;20(1):50–3. https://doi.org/10.1053/j.sempedsurg.2010.10.009.
7. Barthlen W. Surgery in congenital hyperinsulinism - tips and tricks not only for surgeons. A practical guide. Semin Pediatr Surg. 2011;20(1):56–9. https://doi.org/10.1053/j.sempedsurg.2010.10.002.

# Part V
# Common Problems of Infancy and Childhood

# Inguinal Hernia/Hydrocele

**Christie Buonpane and Alfred P. Kennedy Jr**

**Key Points**

1. Diagnosis of an inguinal hernia in children can be made by history and physical examination alone.
2. Due to the increased risk of incarceration, some authors recommend repair for premature infants prior to discharge from the NICU.
3. Premature infants requiring inguinal herniorrhaphy prior to gestational age approximately (institution specific) 56 weeks post conception require at least 12 h of observation after a general anesthetic for concerns of apnea. Full-term infants require postoperative observation if less than 44–46 weeks post conception.
4. Hydroceles that fail to resolve by approximately 12 months of age or vacillate in size deserve groin exploration for concern of a patent processes vaginalis.

Inguinal hernia and hydrocele are the most common problem in children requiring operative correction. Hernia and communicating hydrocele are abnormalities along the same spectrum of incomplete closure of the processes vaginalis after testicular descent or along round ligament.

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1. Types of groin hernias:
   - A. Indirect hernia: Formed by patent processes vaginalis (PPV) and exits lateral to the deep epigastric vessels. This form of hernia is most common in children (“congenital inguinal hernia”).
   - B. Direct hernia: Represents a defect within the inguinal floor (transversalis fascia), medial to the epigastric vessels (“acquired inguinal hernia” or “rupture”).
   - C. Femoral hernia: Also represent a defect in the posterior inguinal floor. The hernia passes through the femoral ring deep to the inguinal ligament and emerges within the femoral canal at the fossa ovalis.
   - D. Sliding hernia: Any hernia of which a portion of the sac consists of viscera.
   - E. Amyand hernia: The vermiform appendix is within the hernia sac.
   - F. Littre’s hernia: A Meckel’s diverticulum is the herniated viscus.
   - G. Richter’s hernia: A portion of the intestinal wall is included within the hernia sac.
   - H. Incarcerated hernia: Any hernia in which a portion of viscera is entrapped and represents a surgical urgency. Incarcerated hernia that are irreducible progress to strangulation.
   - I. Strangulated hernia: Any hernia in which an incarcerated portion of viscera has lost its blood supply and represents a surgical emergency (Figs. 1 and 2).

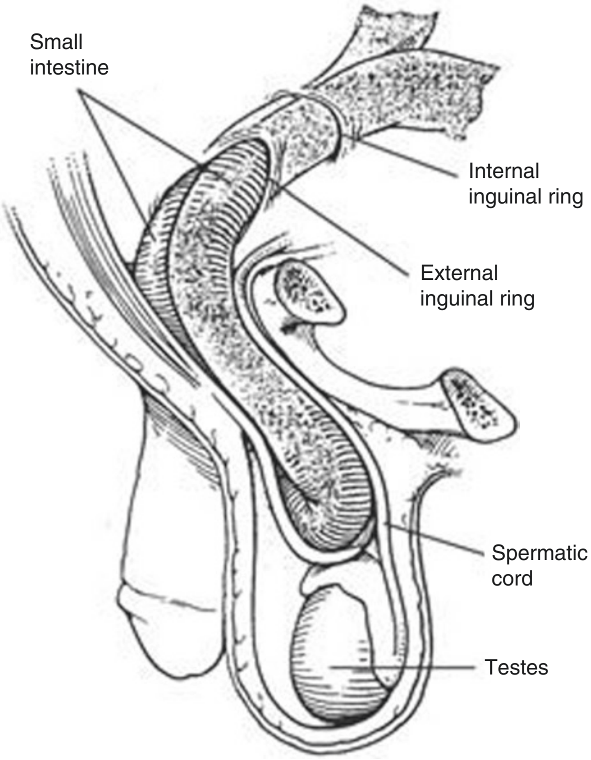

**Fig. 1** Indirect inguinal hernia. The indirect inguinal hernia passes first through the internal inguinal ring and then through the external inguinal ring after traversing the inguinal canal (Source: National Digestive Diseases Information Clearinghouse, National Institutes of Health, Bethesda, MD. http://digestive.niddk.nih.gov/diseases/pubs/dictionary/e-k.aspx)

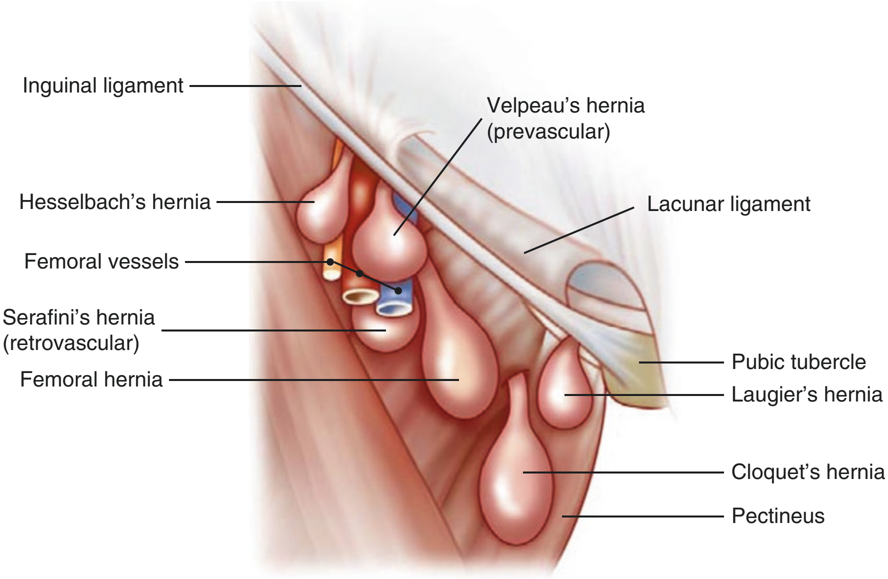
Inguinal ligament
Velpeau's hernia (prevascular)
Hesselbach's hernia
Lacunar ligament
Femoral vessels
Serafini's hernia (retrovascular)
Pubic tubercle
Femoral hernia
Laugier's hernia
Cloquet's hernia
Pectineus

**Fig. 2** Femoral hernia types. Femoral hernias are defects in the abdominal wall between the inguinal ligament and the upper pubic ramus (Source: Dr. Kumar Nishant, under Creative Commons. Attribution Share-Alike 3.0 Unsupported license, from https://upload.wikimedia.org/wikipedia/commons/6/6e/Femoral_hernia_types.jpg)

2. Anatomy:
   - A. Fascial layers of the abdominal wall contribute to layers of the testis and spermatic cord during testicular descent.
     - i. Transversalis fascia → internal spermatic fascia.
     - ii. Internal oblique → cremaster.
     - iii. External oblique → external spermatic fascia.
     - iv. Processes vaginalis (peritoneum) → tunica vaginalis.
   - B. Inguinal canal in children:
     - i. Shorter in infants than adults as the external and internal rings are nearly effaced, which leads to increased risk of incarceration.
     - ii. Scarpa's fascia well developed in children and may be mistaken for external oblique aponeurosis.
3. Embryology:
   - A. Gonads form at fifth week gestation within the retroperitoneal nephrogenic ridges.
   - B. Testis descends to internal ring at 7 months gestation.
   - C. Testis descent mediated by calcitonin gene-related peptide (CGRP) released from the genitofemoral nerve.

D. Descent of testis proceeds along gubernaculum and "drags" a diverticulum of peritoneum known as the processes vaginalis.
E. Obliteration of processes vaginalis mediated by CGRP.
F. Failure of this obliteration leads to the potential for indirect inguinal hernia formation.
G. Ovarian descent arrests within the pelvis.
H. Left testicular descent precedes right side; therefore, right-sided hernias are more common as the likelihood of PPV closure decreases after birth.

4. Incidence:

A. Undescended testis and PPV are more common in prematurity.
B. Approximately 1 in 20 males will develop an inguinal hernia.
C. Family history is positive in approximately 10%.
D. Incidence of inguinal herniation increases with prematurity and is estimated within 10–30%.
E. Incidence of contralateral occult hernia (bilateral hernias) is approximately 10%. This percentage is undoubtedly higher in premature infants and females.

5. Associated conditions:

A. Prematurity.
B. Cryptorchidism (90% have PPV).
C. Connective tissue disorders.
D. Cystic fibrosis.
E. Ascites.
F. Abdominal wall defect.
G. Myelomeningoocele.
H. Ambiguous genitalia.
I. Any condition that increases abdominal pressure will likewise increase the chance of a clinical hernia developing, e.g. chronic lung disease, ventriculoperitoneal shunt, and peritoneal dialysis catheter.

6. Clinical presentation:

A. Asymptomatic inguinal and/or scrotal bulge most common and usually discovered by a caregiver.
B. Lifetime incidence of inguinal hernias approaches 5% in males.
C. Inguinal hernia are 5–10 times as prevalent in males.
D. Sixty percent present on the right side, 25% on the left, 10% bilateral.
E. Diagnosis is clinical and relies squarely on physical examination.
F. Maneuvers to assist in appreciation include an upright examination and Valsalva maneuver.
G. "Silk glove sign" involves the sensation of silk rubbing together or a thickened cord upon palpation of the spermatic cord.

- H. Radiography/ultrasonography generally unnecessary. A “negative” groin ultrasound in no way precludes the presence of a hernia and a false positive is possible.
- I. A “negative” physical examination should prompt caution in groin exploration.
- J. May repeat physical examination at a later visit.
- K. Treatment of a PPV found during laparoscopy for another condition (e.g. appendicitis) should be deferred until clinical findings are apparent.
- L. Within the female population, a mass is usually found within the labia majora. This often represents an ipsilateral sliding ovary and is the size and shape of an almond.
- M. Incarcerated inguinal hernias present as an inguinoscrotal mass, which may be erythematous and tender, are difficult to reduce, and often require sedation to do so. The technique involves firm and continuous pressure along the axis of the spermatic cord while stabilizing the ipsilateral testicle. The examiner will note a sudden “pop” or “gurgle” as the hernia contents return to the peritoneal cavity.
- N. Strangulated inguinal hernias present with signs of bowel obstruction and/or peritonitis. No attempt should be made to reduce the hernia in this situation. Rather, immediate surgical intervention is required.

7. Diagnosis:
   - A. Diagnosis is made via detection of the hernia on physical examination.
   - B. Parents’ description of a bulge that comes and goes at the external inguinal ring is sufficient to merit operative repair.
   - C. Some conditions may mimic hernia.
     - i. Retractile testicle (able to be displaced into scrotum).
     - ii. Lipoma of the cord (empty internal ring).
     - iii. Hydrocele (fluid-filled sac will transilluminate with a light).
     - iv. Lymphadenopathy (located below inguinal ligament).
     - v. Neoplasia (testicular mass).
8. Treatment:
   - A. Operative management:
     - i. Usually performed under general anesthesia.
     - ii. May consider spinal anesthetic for premature infants with chronic lung disease to avoid intubation and subsequent prolonged ventilation. Although the incidence of apnea and bradycardia after either form of anesthetic is probably not significantly different.
   - B. Postoperative monitoring:
     - i. Full-term infants greater than 44 weeks post conception do not require overnight post-anesthetic apnea monitoring.

    ii. Risk of apnea and bradycardia increases with prematurity and concomitant lung disease. Infants with a history of prematurity and/or apnea and bradycardia should receive at least 12 h of monitoring if their post conceptual age is less than 56 weeks.

  C. Timing of operative hernia repair:

    i. Due to the increased risk of incarceration, premature infants should have their hernia repaired prior to discharge from the NICU, particularly if hernia reduction has been necessary.
    ii. Some authors recommend repair outside the neonatal period due to concern over potential neurotoxicity of inhalational anesthetics. Additionally, there may be a lower recurrence rate after herniorrhaphy in older infants. For in-depth discussion related to anesthesia-related neurotoxicity (GAS Trial, PANDA Trial), the reader in referred to the Anesthesia chapter within this textbook.
    iii. For outpatients, delay repair until child is at approximately 56 gestational weeks of age may lessen the risk of apnea after general anesthesia.
    iv. If symptomatic, repair hernia when detected, but plan overnight monitoring in hospital if <56 gestational weeks old (institution specific).

9. Types of repair:

  A. Indirect inguinal hernia:

    i. High ligation of sac at the level of the internal inguinal ring (Marcy repair).
    ii. Inguinal incision within Hesselbach's triangle.
    iii. Incision of external oblique aponeurosis.
    iv. Exposure of spermatic cord.
    v. Exploration of spermatic cord.
    vi. High ligation of indirect inguinal hernia.

  B. Direct inguinal hernia:

    i. McVay or Cooper's ligament repair (approximation of conjoined tendon to ileopubic tract).
    ii. Check for concomitant indirect (Pantaloon) hernia.
    iii. Consider laparoscopic mesh (Lichtenstein) repair in adolescents.

  C. Femoral hernia: similar to direct inguinal hernia repair.
  D. Incarcerated hernia:

    i. If able to reduce, then patient should be admitted for observation of recurrence and subsequent repair within 24–48 h.
    ii. In the rare event that the hernia is non-reducible, emergent exploration and repair are required with concern for strangulation.
    iii. Presence of blood or enteric contents within the hernia sac (strangulation) mandates laparoscopy or laparotomy via La Roque incision.

- iv. Follow patient yearly for signs of recurrence or testicular atrophy secondary to ischemia.
- v. Minimally invasive techniques:
  - i. Intraperitoneal approaches (intracorporeal suturing): laparoscopic approach wherein the hernia sac is ligated, with care to avoid injury to the vas deferens and the testicular vessels, followed by intracorporeal placement of a purse string suture to close the peritoneum and thus obliterate the sac opening.
  - ii. Extraperitoneal approach (suture through the abdominal wall): involves using various types of needles under laparoscopic guidance to percutaneously secure a suture circumferentially around the internal ring, thus obliterating the sac opening.
  - iii. Ultrasound guided (females only): the inguinal hernia is manually reduced under ultrasound imaging and a hollow needle is passed through the skin on the lateral side of the hernia, under and around the hernia and back out the medial side. A silk suture is passed through the needle and the needle is removed. The needle is again inserted, this time passing superficial to the hernia. The same suture is passed through the need and the needle is removed (medial to lateral). The suture now forms a loop circumferentially around the hernia defect and is tied down. A needle laparoscope may be used to confirm obliteration of the hernia defect.
- vi. Contralateral exploration:
  - i. The incidence of contralateral processes vaginalis patency ranges from 20 to 66%.
  - ii. Incidence differs by gender and gestational age (highest incidence in term and preterm female infants with left-sided hernias).
  - iii. Complications from contralateral exploration are reported from 2 to 16% including wound infection, hematoma, injury to spermatic cord structures, injury to intraperitoneal structures, and injury to epigastric or femoral vessels.
  - iv. No consensus recommendation exists for when to consider contralateral exploration. Some practitioners routinely explore the contralateral groin in females and premature infants.
  - v. Peritoneoscopy (70° scope) through the inguinal hernia sac during open repair as well as laparoscopy at the time of laparoscopic repair can assess for a patent processes vaginalis.

10. Hydrocele:
    - A. Represent similar pathology as an indirect hernia, i.e. PPV.
    - B. Asymptomatic hydroceles found in a newborn may be observed.
    - C. Those that fail to resolve by 1 year or are noted to vacillate in size (communicating) deserve formal groin exploration.

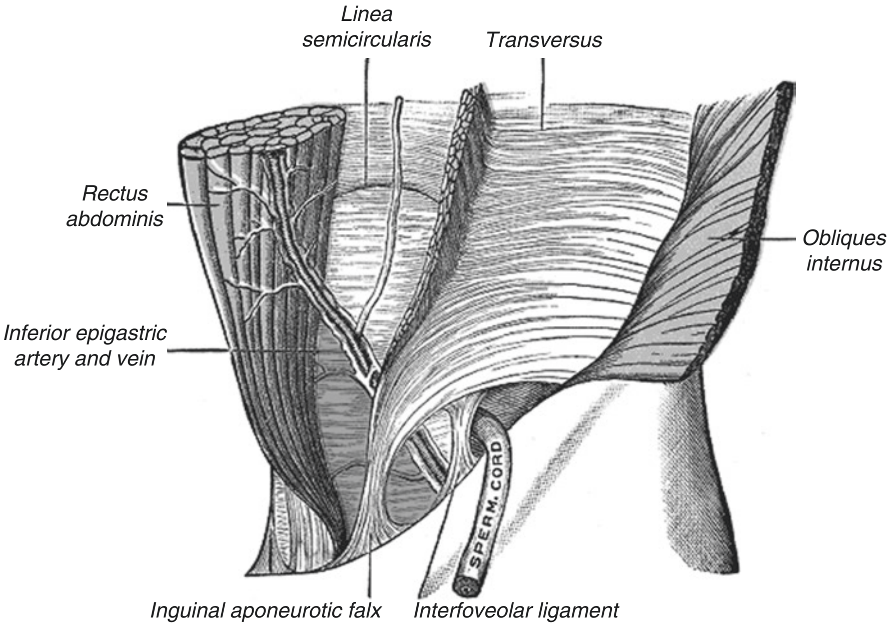
Linea semicircularis
Transversus
Rectus abdominis
Obliques internus
Inferior epigastric artery and vein
SPERM. CORD
Inguinal aponeurotic falx
Interfoveolar ligament

**Fig. 3** Hesselbach's triangle from the outside. Hesselbach's triangle is bounded by the lateral margin of the rectus muscle, the inferior epigastric vessels, and the inguinal ligament (Source: Gray's Anatomy of the Human Body, 20th U.S. ed., 1918, via public domain reproduction rights)

- D. Complete resection of the hydrocele is not necessary and may increase the risk of cord injury. A bottle neck repair suturing the hydrocele sac behind the cord structures is done to prevent recurrence.
- E. "Hydroceles" that arise in adolescents mandate testicular ultrasound to evaluate for associated lesions such as testicular neoplasms (Figs. 3 and 4).

11. Outcomes:
    - A. Injury to spermatic cord:
        - i. Incidence approximates 1/1000 groin explorations.
        - ii. If recognized, perform direct repair of vas deferens with 8-0 absorbable suture, preferably using a microscope.
    - B. Absent vas deferens:
        - i. Consider associated renal abnormality (agenesis) if unilateral.
        - ii. Consider cystic fibrosis if bilateral or atrophic.
12. Disorders of androgen insenstivity:
    - A. Suspicion arises when a "testicle" or "epididymis" is found during exploration of the female groin, usually as a sliding indirect hernia.

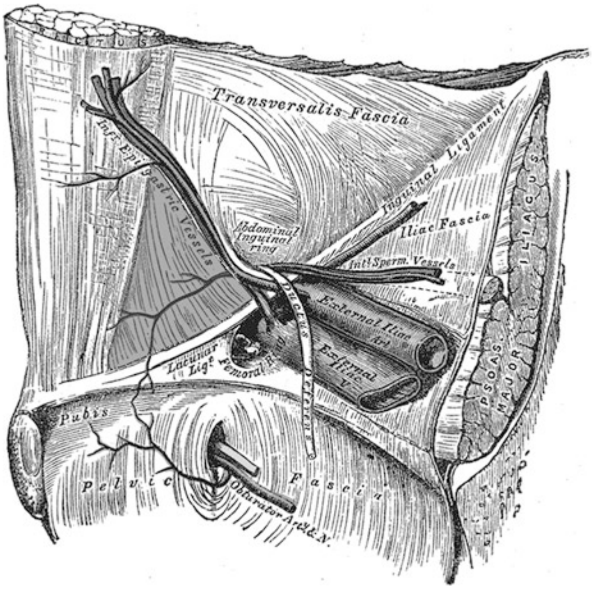
RECTUS
Transversalis Fascia
Inguinal Ligament
Infr. Epigastric Vessels
Abdominal Inguinal ring
Iliac Fascia
Intl. Sperm. Vessels
External Iliac Art.
External Iliac V.
Ductus deferens
Lacunar Ligt.
Femoral R.
ILIACUS
PSOAS MAJOR
Pubis
Pelvic Fascia
Obturator Art. & N.

**Fig. 4** Hesselbach's triangle from the inside. This is the view seen from the laparoscopic approach (Source: Gray's Anatomy of the Human Body, 20th U.S. ed., 1918, via public domain reproduction rights)

B. Consider laparoscopy and/or vaginoscopy to evaluate Mullerian structures (uterus, fallopian tubes, and cervix).
C. Transverse biopsy of the gonad should be performed.

## Additional Notes

Inguinal hernias (from the Greek, "hernios," meaning bud) have been documented from the dawn of civilization. Galen described its predecessor, the patent processes vaginalis (PPV) within the second century AD. Pediatric inguinal hernias today remain one of the most common conditions treated by pediatric surgeons and urologists. Inguinal hernias are almost exclusively indirect in the pediatric population. Although common, surgeons should be able to differentiate the various pathologies that present within the groin to prevent pitfalls in diagnosis. Familiarization with local anatomy and surgical repair will prevent intraoperative and postoperative complication. Inguinal hernia repair for healthy children or term infants is an outpatient procedure, usually performed under general anesthesia. Alternatives include regional (spinal) anesthetic for infants at risk for apnea. The concern for neuroprotective strategies has led to a resurgence of regional anesthetics. Postoperative apnea monitoring originally was reserved for infants younger than 60 weeks post conception. Most authors now recommend overnight monitoring in high-risk patients (prematurity, chronic lung disease) only, although the risk appears to be low. Communicating hydroceles are similar to inguinal hernia with respect to anatomy and hence should be treated as such with the exception of timing of surgery. Incarcerated hernias remain peculiar to the premature and younger patient but may occur at any age. They are true surgical emergencies. Other forms of hernias remain rare within the inguinofemoral region. Complications also remain infrequent after inguinal hernia repair. Recurrence of hernia is unusual (2%) and is more common following repair for incarceration or disease states that increase intrabdominal pressure (ventriculoperitoneal shunt, chronic lung disease of prematurity). Injury to the spermatic cord (<1%) should be a rare event if performed by an experienced pediatric surgeon or urologist. Iatrogenic cryptorchidism is another rare event. Chronic inguinal pain (inguinodynia) is more commonly reported after adult hernia repairs with separate surgical techniques. Mortality directly related to inguinal herniorrhaphy is exceedingly rare.

## Study Questions

1. If you cannot find the vas deferens during a hernia repair, the most likely explanation is:

   a) Iatrogenic injury of the vas deferens
   b) Cystic fibrosis
   c) Disorder of sexual development
   d) Poor surgical technique

Answer: b. Cystic fibrosis is associated with bilateral anatomic variants of the vas deferens.

2. At what age post conception is the risk of postoperative apnea less than 1%?

   a) 44 weeks
   b) 52 weeks
   c) 56 weeks
   d) 58 weeks

Answer: c. 56 weeks. If under this age, patient should be kept for apnea monitoring for at least 12 h postoperatively.

3. An 8-month-old male with unilateral scrotal swelling that does not change in size is brought to your clinic. The scrotum transilluminates on examination and cannot be reduced. What is the treatment?

   a) Emergent surgical repair
   b) Urgent surgical repair
   c) Elective surgical repair
   d) Observation

Answer: d. Observation. Most non-communicating hydroceles will resolve by 1 year of age. If persists, recommend surgical repair.

## Further Reading

Abdulhai S, Glenn I, Ponsky T. Inguinal hernia. Clin Perinatol. 2017;44:865–77.

American Pediatric Surgical Association. Inguinal hernia. Pediatric Surgery Not A Textbook (NaT). https://www.pedsurglibrary.com/apsa

American Pediatric Surgical Association. Inguinal hernia and hydrocele repair. Pediatric Surgery Not A Textbook (NaT). https://www.pedsurglibrary.com/apsa

Holcomb and Ashcraft's Pediatric Surgery, 7th Ed. Elsevier Pub. 2020, Chapter 50 on Inguinal Hernia, pp. 784–800.

Jessula S, Davies D. Evidence supporting laparoscopic hernia repair in children. Curr Opin. 2018;30:405–10.

Kim AG, Jarboe MD. Inguinal and other hernias. Adv Pediatr. 2020;67:131–43.

Pediatric Surgery, 7th ed. CH. 76: Inguinal Hernias and Hydroceles.

Skandalakis JE, Gray SW, Rowe JS. Anatomical complications in general surgery. New York: McGraw-Hill; 1983.

# Umbilical Hernia and Other Disorders of the Umbilicus

**Christie Buonpane and Alfred P. Kennedy Jr**

**Key Points**
1. Umbilical hernias in children often spontaneously close with time.
2. Indications for surgery include failure to close by age 3–5 years, incarceration, strangulation, perforation, or evisceration.
3. Other disorders of the umbilicus can occur including umbilical granulomas, fistulas, sinuses, infections, cysts, and more.

I. Diagnosis and Treatment:

Umbilical hernia is a common abdominal abnormality identified in infancy and is usually self-limiting. Umbilical hernias occur as a result of failure of the umbilical ring to close completely after birth. Only hernias that are very large, symptomatic, or persist past age 3–5 years (surgeon specific) will require operative repair.

Physical examination is sufficient to establish the diagnosis of an umbilical hernia. Most are asymptomatic and are only identified by visualizing a bulge at the umbilicus. The bulge may enlarge with straining and crying. Often, small bowel and/or omentum typically fill the hernia sac. Most are easily reducible. The defects are often small (<1.5 cm) and there may be redundant skin overlying the umbilicus.

Umbilical defects can be identified at birth and about 80% will spontaneously close. If the hernia has not spontaneously resolved, many consider hernia

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repair at age 3–5 years. Hernias with incarceration, strangulation, perforation, or evisceration should receive prompt operative intervention. Incarceration is less common with umbilical hernias as compared to groin hernias.

II. Pathophysiology:

The umbilicus is an essential structure of fetal development. Five structures course the umbilicus during gestation: 3 vessels (2 arteries and 1 vein); the urachus which communicates to the bladder and the omphalomesenteric duct which communicates with the terminal ileum.

Umbilical hernias result from failure of closure of the fascial ring through which the umbilical cord passes. This opening typically closes within a few weeks after the umbilical cord separates.

III. Epidemiology:

Umbilical hernias are more common in African-American, premature, low birth weight, and male children. Also associated with trisomy 21, congenital hypothyroidism, mucopolysaccharidosis, Beckwith-Wiedemann syndrome, and patients requiring peritoneal dialysis.

IV. Surgical Repair:

Repair is done with general anesthesia. The cosmetic result of repair is an important aspect of the surgical procedure. A curvilinear incision is made below the umbilicus, and the hernia sac is dissected away from the fascia. The fascia is closed using either absorbable or nonabsorbable interrupted sutures. The skin is closed with absorbable suture. Large proboscis-type hernias may benefit from flap skin closures such as the 3-flap umbilicoplasty.

V. Complications:

Postoperative complications include bleeding, infection, and recurrence.

VI. Other Umbilical Abnormalities:

a. Delayed separation of the cord
   - Normally separates within 1–2 weeks.
   - A delay in its drop may be associated with a leukocyte adhesion deficiency, a rare immunologic disorder.

b. Umbilical granuloma
   - After cord separation, a small area of granulation tissue may develop.
   - Treated with silver nitrate until the area epithelializes.

c. Umbilical Infections
   - Higher incidence in undeveloped countries.
   - Hand washing and cord care reduce the incidence.
   - Omphalitis is a rare yet aggressive bacterial infection of the newborn that may spread to surrounding tissues. This is a true surgical emergency that may require repeated debridements along with broad-spectrum intrave-

nous antibiotics. The condition may progress to sepsis and death if not recognized early.

d. Omphalomesenteric Remnants

- Remnants of the vitelline or omphalomesenteric duct—include fistulas, sinus tracts, cysts, mucosal remnants, and congenital bands.
- Vitelline fistula—fistula communicates with the ileum manifested clinically by discharge of fecal contents at the umbilicus.
- Vitelline sinus—blind sinus opening at the umbilicus caused by failure of the obliteration of the distal portion of the vitelline duct. Clinically manifests as discharge of mucous at the umbilicus.
- Vitelline cyst—failure of the obliteration of the middle portion of the vitelline duct. The cyst can get infected and/or rupture at the umbilicus, with the discharge of purulent effluent.
- Vitelline band—failure of the obliteration of the vitelline duct leading to a fibrous band that may lead to intestinal volvulus, obstruction, and/or strangulation.

e. Urachal Remnants

- Urachal fistula—clear drainage from the umbilicus due to a fistula connecting the umbilicus with the apex of the urinary bladder. The etiology is related to incomplete urachal obliteration. Diagnosis is usually made with ultrasonography with subsequent surgical excision to prevent complications including infection and metachronous malignancy.
- Urachal cysts—often cause infection and present as a painful mass between the umbilicus and suprapubic region also related to failure of obliteration of the middle portion of the urachus. Treatment is similar to the fistula.

Umbilical anomalies are a common problem seen by every pediatric surgeon. They have been described in surgical textbooks dating back to the early twentieth century. The appearance of the umbilicus was recognized in early works which recommended observation and compression. Most umbilical hernias resolve without intervention. Those that persist beyond the third birthday are likely to require operative closure. Large umbilical defects (greater than 1.5 cm) are unlikely to resolve and earlier intervention may be indicated. Little has changed in the surgical management of umbilical hernias over the past 100 plus years. An understanding of the anatomic structures which course the umbilicus in fetal life helps determine other considerations such as associated anomalies including urachal and omphalomesenteric remnants. Urachal and vitelline remnants often present with infection and thus require surgical excision (Fig. 1).

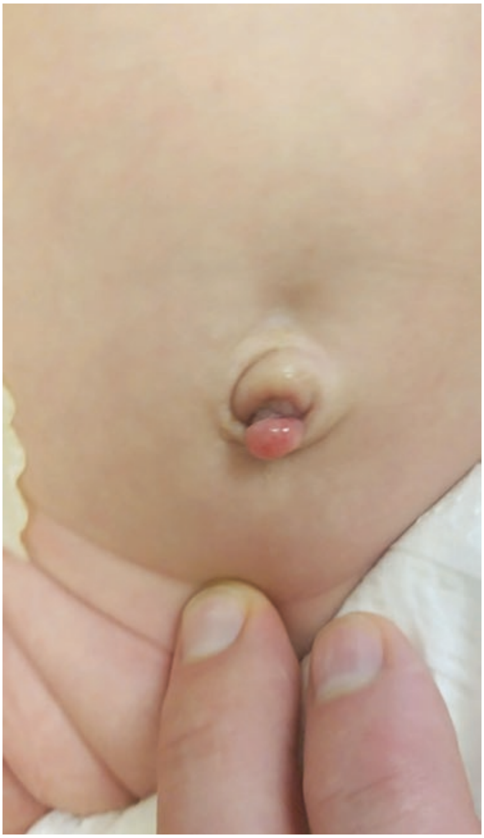

**Fig. 1** Umbilical granuloma. Source: Alfred P. Kennedy, Jr., MD

## Study Questions

1. A 14-month-old female presents with a 1.5-cm reducible umbilical hernia. The parents describe that the hernia increases in size with crying. What is the treatment?
    a) Elective repair
    b) Abdominal binder
    c) Reassurance to parents
    d) Emergent repair

Answer: c) Reassurance to parents. The patient is below 3 years of age, the hernia is asymptomatic and there is no indication for surgical intervention at this time.

2. A 9-month-old male presents with clear umbilical drainage. What is the most likely diagnosis?
    a) Umbilical hernia
    b) Omphalitis

c) Umbilical granuloma
d) Urachal remnant

Answer: d) Urachal remnant. Clear drainage is typically associated with a urachal remnant.

## Further Reading

American Pediatric Surgical Association. Umbilical disorders. Pediatric Surgery Not a Textbook (NaT).

Hegazy A. Anatomy and embryology of umbilicus in newborns: a review and clinical correlations. Front Med. 2016;10(3):271–7.

Holcomb and Ashcraft's pediatric surgery, 7th Ed., Holcomb et al. Eds., Elsevier. 2020, Chapter 49: Umbilical and other abdominal wall disorders, pp. 780–783.

Pediatric surgery, 7th ed., Coran et al Eds., Elsevier. 2012, Chapter 74: Disorders of the Umbilicus, pp. 962–972.

Takasu H, Watanabe Y. Umbilicoplasty with 3 triangular skin flaps and excised diamond-shaped skin flap. J Pediatr Surg. 2010;45(10):2041–4.

Zens T, Nichol P, Cartmill R, Kohler J. Management of asymptomatic pediatric umbilical hernias: a systematic review. JPS. 2017;52:1723–31.

# Intussusception

**Roland J. Scorpio**

**Key Points**

1. Idiopathic intussusception represents 90% of all cases.
2. Hypertrophy of Peyer's patches is often present in conjunction with a pre-existing viral infection.
3. Colicky abdominal pain is always present.
4. Ultrasound is the diagnostic study of choice.
5. Contrast or air enema is highly successful in reducing the intussusception.

Intussusception is the invagination of a segment of intestine into another segment of intestine (Fig. 1). Idiopathic ileocolonic intussusception occurs most commonly in children aged 6 months to 2 years old. Most cases can be reduced with hydrostatic or air enema; few cases will need operative repair. Ileocolonic intussusception past the age of 2 years old brings up the possibility of a pathologic lead point. Small bowel into small-bowel intussusception occurs frequently in children of all ages and the usual course is spontaneous resolution. Occasionally small bowel to small-bowel intussusceptions are due to pathologic conditions such as cystic fibrosis, Henoch–Schonlein Purpura (HSP), polyps, or tumors (Fig. 2).

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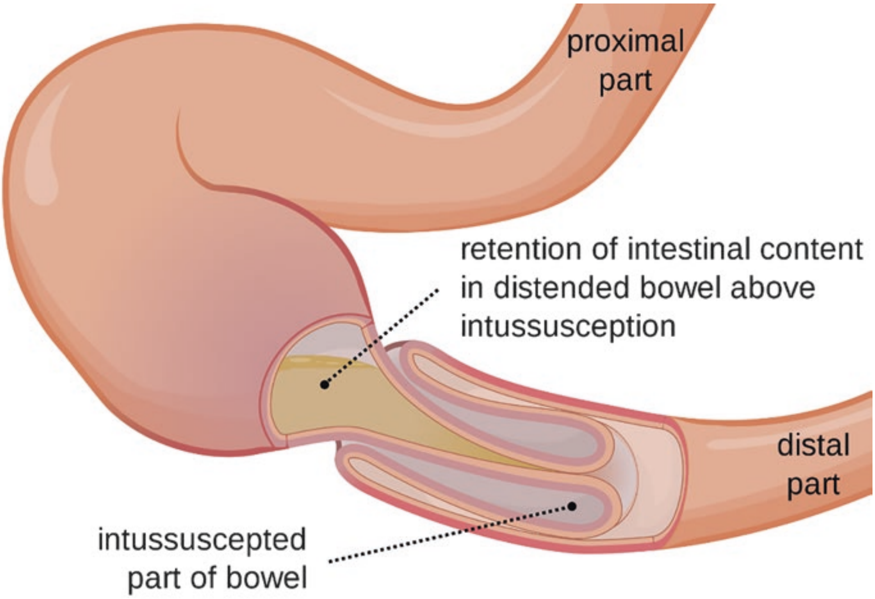
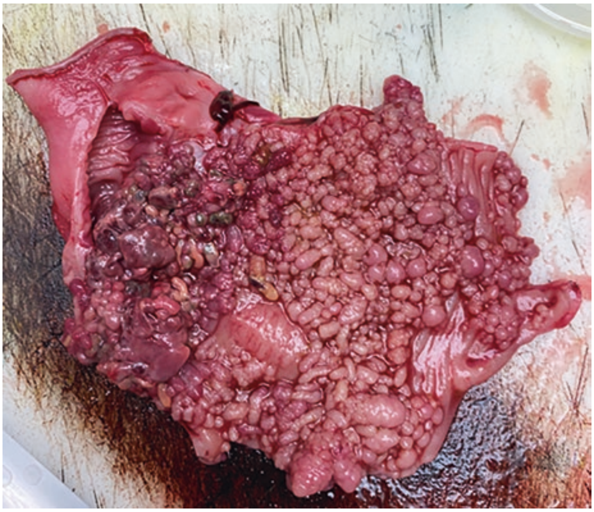

proximal part
retention of intestinal content in distended bowel above intussusception
distal part
intussuscepted part of bowel

**Fig. 1** Intussusception of proximal bowel into distal bowel causing obstruction (Source: Aleksander P. Remesz, Creative Commons Share-Alike Unported 3.0 license. https://commons.wikimedia.org/wiki/File:Intussusception_EN.svg, Accessed 12 Nov 2021)

**Fig. 2** Pathologic specimen of resected jejuno-jejunal intussusception caused by juvenile polyposis acting as a pathologic lead point (Source: Marc Lessin, MD)



1. Etiology:
    a. Idiopathic: 90% caused by virus, hypertrophy of Peyer's patches
        i. No pathologic lead point

    - ii. Often presumed to be secondary to virus
        1. Retrovirus
        2. Reoviruses
        3. Echoviruses
    - iii. Seasonal variation
        1. Increased in spring/summer
        2. Increased in middle of winter
  - b. Organic/pathologic: 7–10% due to Meckel's diverticulum, polyps, tumor, cysts, Henoch–Schonlein Purpura (HSP). HSP results in intussusception because edematous vasculitic mucosa becomes a pathologic lead point [1].
  - c. Postoperative: <3%

2. Location:
   - a. Ileocolic: 90%
   - b. Ileo-ileal: 5% (will usually resolve spontaneously and does not require treatment by a surgeon)
   - c. Other (jejuno-ileal; jejuno-jejunal): 5% (Fig. 1)

3. Incidence in US
   - a. 1.9–4 per 1000 live births
   - b. Male-to-female ratio: 3 to 2

4. Pathophysiology:
   - a. The segment of bowel that enters the distal lumen is called the intussusceptum.
   - b. The distal segment of bowel filled by the intussusceptum is called the intussuscipiens.
   - c. Venous compression occurs from the invagination of the bowel resulting in venous stasis and subsequent decreased arterial flow.
   - d. Signs and symptoms of bowel obstruction
   - e. Bowel necrosis
   - f. Perforation
   - g. Sepsis
   - h. Shock, if untreated which can be fatal in 2–4 days

5. Clinical findings: One-third of children with intussusception have the triad of intermittent abdominal pain, palpable abdominal mass, and currant jelly stools.
   - a. Symptoms [2]
     - i. Colicky abdominal pain, 78%
     - ii. Repeated bouts of vomiting, 95%
     - iii. Currant jelly stools, <25% of cases [3]
     - iv. Passage of bloody mucus, rectally

- b. Signs
    - i. Sausage-shaped mass in right upper abdomen
    - ii. Emptiness in the right lower quadrant (RLQ) (Dance's sign)
    - iii. Fever is common

6. Differential diagnosis
    - a. Gastroenteritis
    - b. Acute appendicitis
    - c. Small-bowel obstruction
    - d. Meckel's diverticulitis

7. Diagnosis
    - a. History and physical (as described above)
    - b. Laboratory studies
        - i. Complete blood count: leukocytosis
        - ii. Electrolytes: dehydration, electrolyte disturbances
        - iii. Blood in stool
    - c. Radiology
        - i. Abdominal X-ray:
            1. Usually unremarkable but may show stool in ascending colon (diagnosis less likely), absence of air in RLQ, soft tissue mass in right upper quadrant (present 60% of times), and evidence of small-bowel obstruction.
        - ii. Ultrasound
            1. Sensitivity 98%, specificity 98%
            2. Positive predictive value 87%
            3. Negative predictive value 100%
            4. Target sign – intussusception is larger than mean diameter of small bowel giving a target appearance
            5. Pseudo kidney sign – seen on longitudinal scans, the thickened intussusceptum bowel wall mimics a kidney.
        - iii. Air or water-soluble enema (Fig. 3)
            1. Can identify and reduce the intussusception.
            2. Rule of 3 s: Hold column of barium 3 feet high for 3 min, drain in between attempts, and 3 total attempts.
        - iv. Abdominal CT and MRI (sometimes performed when considering alternate diagnoses)
            1. Not first-line imaging modalities
            2. Demonstrates the intussusception
            3. Demonstrates lead point

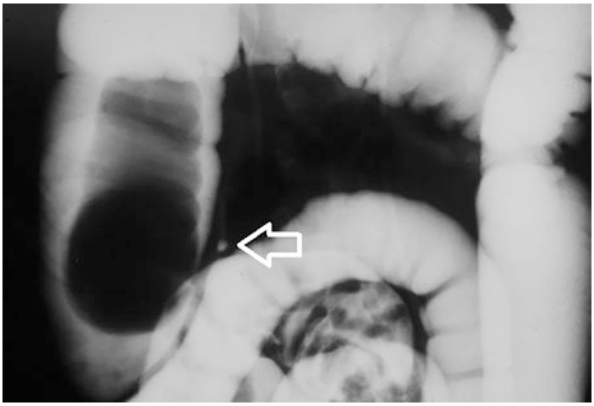


**Fig. 3** Water-soluble contrast enema in ileocolic Intussusception: filling defect in cecum. Note the appendix (arrow) has filled (Source: Marc Lessin)

8. Treatment
    a. Nasogastric tube decompression if signs of obstruction
    b. Intravenous fluids
    c. Antibiotics if necrotic bowel, perforation, or sepsis
    d. Fluoroscopically guided reduction with air or water-soluble contrast enema [4]
        i. Successful 85% of times, if no lead point is present
        ii. Partial reduction and patient is stable, may repeat study 2–3 h later [5]. The partial reduction allows for resolution of the bowel enema, facilitating reduction.
        iii. Must show reflux of contrast or air into terminal ileum.
    e. Ultrasound-guided reduction [6]
        i. Used mainly in Eastern Hemisphere
        ii. Advantage no radiation exposure
        iii. Success rate 70–95%
    f. Operative treatment
        i. Indications
            1. Peritonitis or perforation
            2. Unsuccessful hydrostatic reduction
            3. Pathologic lead point
        ii. Technique
            1. Open
                a. Incision: RLQ, transverse, or midline
                b. Manual reduction of intussusception, similar to squeezing toothpaste from a tube
                c. Appendectomy is sometimes performed at same operation but may increase length of stay and cost of treatment [7]

    d. In severe cases, bowel resection may be required
        i. Manual reduction unsuccessful
        ii. Perforation and spillage present
        iii. Necrotic bowel present
2. Laparoscopic
    a. The terminal ileum is gently pulled out from the intussusception into the cecum using atraumatic graspers
    b. Appendectomy is sometimes performed at same operation
3. Resection of lead point if present

## Additional Notes

If intussusception is suspected, then it is imperative the diagnosis be ruled in or out. Intussusception represents a surgical emergency for two reasons. First, the intussusceptum is compressed within the intussuscipiens which leads to vascular compromise of the bowel that will become necrotic and gangrenous if it is untreated for enough time. Second, intussusception represents a bowel obstruction. While obstructed, the child is slowly developing dehydration, electrolyte abnormalities, and hypovolemic shock.

The high rate of reduction with hydrostatic enema means that the majority of children with intussusception are treated without an operation. Air reduction which is practiced at most pediatric centers is slightly more successful than hydrostatic reduction and uses less radiation [8, 9]. As time progresses, the trend is to use air-contrast enema reduction over water-soluble contrast enema reduction. The majority of centers in the US use fluoroscopically guided reduction, but sonographic guided reduction is being demonstrated to be an effective method. In recent years, there is as an increasing trend for uncomplicated intussusception to be treated without hospital admission after reduction. If children are able to tolerate oral intake, they are discharged home from the emergency department after a short period of observation.

## Study Questions

1. A 9-month-old infant presents with colicky abdominal pain. You suspect intussusception, after a complete history and physical examination, your first study is:
    a. Abdominal X-ray
    b. Ultrasound
    c. Abdominal computed tomography
    d. Contrast enema

Answer: (b.) Ultrasound. Ultrasound has the highest positive predictive value in diagnosis of intussusception. The high suspicious of intussusception in this child should make ultrasound the first study. Abdominal X-ray may diagnose a radiographic Dance's sign with a tissue mass in the right upper quadrant, but it is not as sensitive or specific as ultrasound. Abdominal X-ray can help in choice of management when it identifies perforation due to any cause. Computed tomography carries the risk factors of ionizing radiation so should not be the first-choice study. Water-soluble or air-contrast enema may be necessary if the diagnosis of intussusception is confirmed on ultrasound.

2. All of the following are symptoms of intussusception except:
   a. Colicky abdominal pain
   b. Passage of bloody mucus
   c. Vomiting of blood
   d. Currant jelly stools

Answer: (c.) Vomiting of blood. Vomiting and bilious vomiting can be present in intussusception because of the intestinal obstruction; however, vomiting of blood would not be expected.

3. All are true concerning abdominal ultrasound and intussusception except:
   a. Sensitivity 98%
   b. Target sign occurs when the diameter of the small bowel is larger than the intussusception giving a target appearance.
   c. Negative predictive value is 100%.
   d. Pseudo kidney sign is the thickened intussusceptum bowel wall mimicking a kidney.

Answer: (b.) Target sign occurs when the diameter of the small bowel is larger than the intussusception giving a target appearance. This statement is not true of target sign of intussusception seen on ultrasound. The opposite is true: target sign is caused when the diameter of the intussusception is greater than the diameter of the small bowel, not the other way around.

4. Bowel resection is required in all but:
   a. Necrotic bowel
   b. Perforation
   c. Unsuccessful enema reduction
   d. Presence of a lead point

Answer: (c.) Unsuccessful enema reduction. Resection is bowel required for unsuccessful manual reduction during an operation, not for unsuccessful enema reduction, before any operation is performed. Unsuccessful enema reduction is an indication for either another attempt at enema reduction after a period of rest or for proceeding to operative manual reduction of the intussusception.

## References

1. Little KJ, Danzl DF. Intussusception associated with Henoch-Schonlein purpura. J Emerg Med. 1991;9(Suppl 1):29–32.
2. Kupperman N, O'Dea T, Pinckney L, Hoecker C. Predictors of intussusception in young children. Arch Peadiatr Adolesc Med. 2000;154:250–5.
3. Padilla BE, Moses W. Lower gastrointestinal bleeding and intussusception. Surg Clin N Am. 2017;97:173–88.
4. Del-Pozo G, Albillos JC, Tejedor D, Calero R, Rasero M, de-la-Calle U, Lopez-Pacheco U. Intussusception in children: current concepts in diagnosis and enema reduction. Radiographics. 1999;19(2):299–319.
5. Navarro OM, Daneman A, Chae A. Intussusception: the use of delayed, repeated reduction attempts and the management of intussusceptions due to pathologic lead points in pediatric patients. AJR Am J Roentgenol. 2004;182(5):1169–76.
6. Khong PL, Peh, WCG, Lam CHL, Chan KL, Cheng W, Lam WWM, Ai VHG, Saing H, Tam PKH, Leong LLY, Low LCK. Ultrasound-guided hydrostatic reduction of childhood intussusception: Technique and demonstration. RadioGraphics. 2000;20(5). Radiological Society of North America, Oak Brook, LI. doi:https://doi.org/10.1148/radiographics.20.5.g00see11, Accessed 9 Nov 2021.
7. Wang A, Prieto JM, Ward E, Bickler S, Henry M, Kling K, Thangarajah H, Ignacio R Jr. Operative treatment for intussusception: Should an incidental appendectomy be performed? J Pediatr Surg. 2018;54(3):495–9.
8. Beres AL, Baird R. An institutional analysis and systematic review with meta-analysis of pneumatic verses hydrostatic reduction for pediatric intussusception. Surgery. 2013;154(2):328–34.
9. Kaplan SL, Magill D, Felice MA, Edgar JC, Anupindi SA, Zhu X. Intussusception reduction: effect of air vs. liquid enema on radiation dose. Pediatr Radiol. 2017;47(11):1471–6.

# Gastrointestinal Foreign Objects

**Biren Desai and Brian Maksimak**

**Key Points**

1) Any esophageal foreign body causing respiratory compromise requires emergent removal, regardless of NPO status.
2) Removal of an esophageal button battery is considered emergent as both morbidity and mortality quickly rise the longer the battery remains in place. Gastric button batteries require emergent removal if the patient is symptomatic.
3) Multiple ingested high-powered magnets are another foreign body which entails a higher level of morbidity and mortality and should be removed emergently via endoscopy if accessible or laparoscopically if there are concerning symptoms.
4) For ingestions in which conservative management is elective, patients should be carefully monitored for deterioration and should have serial physical examinations +/− imaging to monitor for the foreign body's passage through the GI tract.

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## Introduction

Foreign body ingestions in the pediatric population most often occur in children less than 5 years of age and are typically accidental in nature. Common non-food objects that are ingested include coins, magnets, batteries, and jewelry.

This chapter will first focus on basic commonalities in presentation, evaluation, and diagnostic workup when a patient presents for a foreign body ingestion. Afterwards, specific details regarding the ingestion of button batteries, magnets, coins, sharp items, and food impactions will be discussed.

## Presentation

1) Foreign body ingestions are often witnessed by the patient's family.
2) In some instances, it is unknown or unclear if an ingestion has occurred.
3) Clinical acuity can range from a stable patient to a hemodynamically unstable patient.
4) If perforation has occurred after an ingestion, patients may present with abdominal pain and clinical features consistent with sepsis.

## Evaluation

1) Symptoms to assess: Inability to tolerate secretions, chest pain, stridor, dysphagia, fussiness, vomiting, and hematemesis. Conversely, the patient may be asymptomatic.
2) It is important to determine the following in the history as it may alter timing of endoscopic and/or surgical intervention:
    a) What is the object thought to be? How many objects were ingested? What time did the ingestion occur?
    b) Time of last oral intake.
    c) Approximate length and diameter of the object.
3) Physical examination:
    a) Vitals: Evaluation for fever, tachycardia, tachypnea, hypoxia, hypotension.
    b) Evaluation of the oropharynx for evidence of trauma.
    c) Pulmonary examination evaluating for shortness of breath, unequal breath sounds, and wheezing.

## Additional Information

1) Determining timing of an endoscopic intervention is a critical management decision:
    a) Emergent—performed less than 2 h from time of presentation, regardless of NPO status.
    b) Urgent—performed less than 24 h from time of presentation, following institutional NPO guidelines.
    c) Elective—performed greater than 24 h from time of presentation, following institutional NPO guidelines.
2) Consideration should be given to repeat X-rays immediately prior to an upper endoscopy to ensure the foreign body is still in the esophagus or stomach prior to proceeding.
3) The use of glucagon has mixed data and is generally avoided unless the patient is in a center where upper endoscopy is not offered.

## Radiologic Findings

1) X-ray:
    a) All patients, even those with a radiolucent foreign body, should have an anterior-posterior and lateral chest X-ray +/− abdominal X-rays.
    b) A radiopaque object's location can be determined when comparing the two imaging planes.
    c) May demonstrate air trapping within the lungs and mediastinal air leak.
2) Contrast esophagram:
    a) Can demonstrate perforation or stricture if present.
    b) Can help diagnose fistulas between the esophagus and surrounding structures, such as the trachea.
    c) Often can make foreign body retrieval more difficult by obscuring direct visualization during an upper endoscopy.
3) Computed tomography (CT):
    a) Can be used to identify precise location of the object.
    b) Good imaging modality to detect perforation and air leak.
    c) Negative factor is the significant amount of radiation exposure.

## Button Battery Ingestion

1) A flat circular battery that resembles a coin/button.
2) Mechanism of injury: Generation of hydroxide-free radicals that damage the local mucosa via alkalinization of the pH.
    a) Injury can often begin less than 30 min from time of ingestion.
3) Complications post removal can occur from days to weeks after ingestion and include:
    a) Tracheoesophageal fistulas, esophageal perforation, esophageal stricture formation, vocal cord paralysis, mediastinitis, cardiac arrest, pneumothorax, and aortoenteric fistula.
4) Evaluation pearls:
    a) History—batteries can be found in remote controls, watches, toys, and hearing aids.
    b) Imaging—A "double halo" sign can be seen in the AP CXR and the "step off" sign can be seen in the lateral CXR.
    c) If there is hematemesis anywhere from time of removal to 3 weeks, it is assumed the etiology is an aortoenteric fistula that will require prompt cardiothoracic surgical intervention. There is a high mortality associated with this complication.
5) Management:
    a) Ingestion of a button battery that is present in the esophagus is an emergent indication for endoscopy and should take precedence over almost all other OR cases.
    b) Prior to endoscopy you can give sucralfate 1 g/10 ml (10 mL PO every 10 min, up to 3 doses) or honey (10 mL every 10 min in children >1 year of age) if the battery is located in the esophagus less than 12 h in order to slow tissue damage.
    c) If there is no concern for perforation, irrigating the injured area with 50–150 mL of sterile acetic acid can be performed to neutralize pH.
    d) Timing of removal is more controversial if the battery is located within the stomach:
        i. Gastric button batteries are less likely to cause a severe injury.
        ii. Observation can be considered in the absence of clinical symptoms when the ingestion occurred less than 2 h from time of presentation and if the battery is confirmed to be smaller than 20 mm and/or the child is older than 5 years of age.
6) Care and monitoring post removal:
    a) Immediately after removal of a button battery in the esophagus, the patient should be hospitalized in the PICU to monitor for complications.

    b) If evidence of esophageal injury, patient should be kept NPO, started on IV antibiotics, and further imaging should be pursued in the form of a CT angiography and/or MRI to determine risk for developing an aortoenteric fistula.

        i. If there is inflammation extending to the aorta, preemptive surgical management in the form of a thoracotomy and aortic grafting may be required given the high mortality associated with an aortoenteric fistula.

    c) If no significant injury to aorta or near aorta, an esophagram can be obtained to exclude a perforation.

## Magnet Ingestion

1) Metals (such as neodymium) which can produce a high-powered magnetic attraction.
2) Mechanism of injury: If two magnets are in adjacent loops of bowel, their magnetic fields could attract the other and cause them to come together with the bowel wall in the middle, thus causing the below complications.
3) Complications:
    a) Entero-enteric fistula formation, perforation, peritonitis, and bowel ischemic/necrosis.
4) Evaluation pearls:
    a) History—Determining the number of magnets ingested is critical as most complications require at least two magnets in different areas of the GI tract to occur.
    b) Imaging—Magnets are radiopaque given their metallic composition and can be visualized in the GI tract.
5) Management:
    a) If more than one magnet is ingested, emergent removal should be pursued even if the patient is asymptomatic.
    b) If the magnet has traversed passed the ligament of Treitz, removal with small-bowel enteroscopy should be pursued if possible. Otherwise, the pediatric surgery team should be consulted for possible laparotomy.
    c) Conservative management with laxative therapy and observation can be considered if it is confirmed that only one magnet was ingested.
6) Care and monitoring post removal:
    a) If a conservative approach is elected, the patient should be monitored closely by family as to not have any further metallic or magnetic ingestions.
        i. All stools should be examined for evidence of passage.

## Coin Ingestion

1) Coins are the most common foreign body ingestion in children within the United States.
2) Mechanism of injury: If the coin remains in the same position for prolonged periods, local tissue damage and necrosis can occur.
3) Complications:
    a) Respiratory compromise if the coin obstructs the lumen of the upper oropharynx proximal to the trachea.
    b) If the esophagus becomes completely obstructed, the patient will be unable to manage their oral secretions.
    c) Bowel perforation or obstruction can be seen if the coin is wider than 25 mm in diameter.
        i. The most common locations for coin impaction include the level of the aortic arch, pylorus, duodenal sweep, and ileocecal valve.
4) Evaluation pearls:
    a) History—Determine which coin was ingested to approximate the size. Age of the child is also important as older children are less likely to have an impaction.
    b) Imaging—Attention must be given to the two-view chest X-ray to ensure the coin-like object is not a button battery. Coins will not have the “double halo” or “step off” sign.
5) Management:
    a) All asymptomatic esophageal coins should be removed urgently via upper endoscopy.
    b) All symptomatic (respiratory compromise or inability to manage secretions) esophageal coins should be removed emergently.
    c) If the coin has passed into the stomach or beyond, conservative management can be pursued with +/−laxative therapy.
        i. All stools should be examined for evidence of the passage of the object.
6) Care and monitoring:
    a) If the coin has not passed several days after the ingestion, serial X-rays as an outpatient can be considered until clearance is documented.
    b) Elective retrieval should be scheduled if the coin has not spontaneously passed within 2–4 weeks after ingestion.

## Pointed and Sharp Object Ingestion

1) Many objects that are ingested have sharp edges. The most common ingestions include straight pins, open safety pins, hairbrush bristles, and sewing pin needles. In addition, organic materials such as fish or chicken bones can be a common ingestion.
2) Mechanism of injury: Mucosal injury and perforation as the object travels through the GI tract.
3) Complications:
    a) Sharp objects have the potential to perforate the organ in which they reside with subsequent extraluminal migration and can progress to other disease states like peritonitis and sepsis.
        i. The most common site of perforation is the ileocecal valve.
    b) May result in abscess or fistula formation.
    c) On rare occasions, catastrophic complications can be seen and include rupture of the common carotid artery and aortoenteric fistula.
4) Evaluation pearls:
    a) History—Special attention should be given to chest pain and dysphagia as this may indicate the object is still in the esophagus.
    b) Imaging—Many objects are radiolucent and will not be visualized on standard X-ray, thus a careful history is critical. Consideration can be given to obtain a CT scan, MRI study, ultrasound, or an upper GI barium swallow study.
5) Management:
    a) A sharp object is considered a medical emergency if it is in the esophagus and should be removed with an upper endoscopy.
    b) If X-rays do not demonstrate a radiopaque object, an urgent upper endoscopy should be pursued if there is a high clinical suspicion for ingestion.
    c) Surgical consultation should be obtained if there is evidence of neck swelling, crepitus, peritonitis, or respiratory compromise.
        i. Surgical consultation should also be considered if the sharp object has traversed passed the Ligament of Treitz.
6) Care and monitoring:
    a) If conservative therapy is elected, the patient should be admitted to the hospital for serial examinations and imaging to monitor progress of passage.
    b) If the object has not passed between 72 and 96 h from time of ingestion, surgical removal should be pursued given the low likelihood of spontaneous passage.

## Esophageal Food Impaction

1) Food impaction in a pediatric patient is most often related to underlying eosinophilic esophagitis.
2) Mechanism of injury: Impaction can cause respiratory compromise if proximal to the trachea. Impaction in the distal esophagus can lead to poor oral secretion management.
3) Complications:
   a) Esophageal mucosal injury
   b) Perforation
4) Evaluation pearls:
   a) History—Chest pain and difficulty swallowing are key history points. History of esophageal pathology in the patient and family is also important.
   b) Imaging—Food impactions are more likely to be radiolucent on X-ray. Consideration can be given to obtain computed tomography, magnetic resonance imaging, and ultrasounds.
      i. Upper GI barium swallow should not be performed in cases of food impaction as there is a risk of aspiration if the contrast cannot traverse past the food bolus.
5) Management:
   a) If the food impaction does not self-resolve, urgent upper endoscopy with subsequent esophageal biopsy sampling is indicated to further delineate an underlying etiology.
   b) If there are symptoms of respiratory compromise or inability to tolerate secretions, emergent endoscopy should be pursued.
   c) Typically, a food impaction needs to be removed piecemeal during an upper endoscopy.
      i. Advancing the food impaction into the stomach is also an option but should be avoided if the cause of the impaction is unknown as it can lead to perforation.
6) Care and monitoring:
   a) Patients should undergo a complete evaluation to identify the underlying etiology of the food impaction with subsequent management to prevent re-occurrence.

## Study Questions

1) A 3-year-old child presents to the emergency department 1 h after witnessed ingestion of a button battery. Which of the following statements is INCORRECT?

   a. Emergent removal is required if the battery is in the esophagus.
   b. Honey can be given every 10 min prior to emergent endoscopy if the battery is in the esophagus.
   c. If evidence of esophageal injury at the time of the endoscopy, a CTA or an MRI should be obtained to evaluate for a possible developing aortoenteric fistula.
   d. Removal of the button battery is not indicated if located in the stomach, 25 mm in size and the patient is asymptomatic.

   Answer:

(d) Removal of the button battery is not indicated if it is in the stomach, 25 mm in size and the patient is asymptomatic. Observation for button batteries located in the stomach is only recommended in asymptomatic patients 5 years of age or older and/or a button battery <20 mm in diameter. Due to the size of the battery and age of the patient, removal of the battery and evaluation for esophageal injury should be done in the next 24–48 h or sooner if patient becomes symptomatic.

2) Which clinical presentation would require emergent foreign body removal?

   a. Child with a coin in the lower esophagus who is asymptomatic.
   b. Child with mild abdominal discomfort and a coin in the stomach.
   c. Child with coin in the upper esophagus who is not handling their secretions.
   d. Child who is asymptomatic with a single magnet in the stomach.

   Answer:

(c) Child with a coin in the upper esophagus who is not handling their secretions. Inability to handle secretions with a foreign body in the esophagus requires emergent removal in the OR or endoscopy suite. The other scenarios/foreign bodies can be monitored radiographically or removed urgently or electively if they fail to pass.

3) All the following should be avoided in a patient with an esophageal food impaction except for:

   a. Obtain an esophagram to determine the location of the food bolus.
   b. Trial of glucagon if the option of endoscopic removal is not readily available.
   c. Allowing the patient to drink water to try and pass the food bolus.
   d. Attempting to push the food bolus into the stomach with the endoscope.

Answer:

(b) Trial of glucagon if the option of endoscopic removal is not readily available. An esophagram is not recommended as the contrast can obscure visualization at the time of the scope. There is also the risk of aspiration with an esophagram as well as drinking fluids. Pushing the food bolus into the stomach is not recommended due to the risk of perforation.

## Further Reading

Kramer RE, et al. Management of ingested foreign bodies in children. J Pediatr Gastroenterol Nutr. 2015;60(4):562–74.

National Button Battery Hotline: 1-800-498-8666. https://www.poison.org/articles/button-batteries

# Airway Foreign Objects and Bronchoscopy

**Jeremy Mock**

**Key Points**

1) Foreign body aspiration should be a consideration in any child with new or acute onset choking or cough. It is known as the great mimicker as it can be misdiagnosed as asthma, croup, or pneumonia.
2) Rigid bronchoscopy is the gold standard for diagnosis of airway foreign body and should be pursued if suspicion is high based on history, physical exam, or imaging.
3) Complete airway obstruction from an airway foreign body is an absolute emergency and carries with it high potential for mortality.
4) Parental/caregiver education and prevention are the mainstay for reducing airway foreign bodies.

1. Background
    a. Airway foreign bodies represent a class of avoidable incidents with high predilection for young children and real potential for morbidity and mortality in all age groups. They are a diagnostic challenge and despite a downturn in deaths, incidence of these events remains relatively consistent over the years [1]. Children under 3 years make up the most common demographic stemming from a variety of reasons: tendency of children to explore with their mouth, lack of advanced dentition to allow for complete chewing of food,

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failure to distinguish edible vs inedible objects, and frequent distraction when eating [2]. Imaging assessments are often lacking in diagnostic utility [3], and both history and physical examination can be vague. Endoscopic assessment with laryngoscopy and bronchoscopy (rigid or flexible) is the mainstay of diagnosis, and a high index of suspicion is necessary in any case in which foreign body aspiration is a possibility [1]. Adding to this, the heterogeneity of objects ingested creates a myriad of considerations in terms retrieval of the object. Ultimately, communication and coordination among all specialties involved are critical to timely identification of these patients and successful therapeutic intervention.

2. Epidemiology
    a. Airway foreign bodies can occur at any age, but most commonly occur in infants and toddlers. Under 5 years of age accounts for 84% of cases [1].
    b. There is male predominance with male-to-female ratio of 1.4:1 [2].
    c. Fourth leading cause of accidental fatalities in children under 3 years and accounts for 7% of all pre-hospital mortalities in this age group [4].
    d. Types of foreign body display global variation based on diet and cultural trends.
        i. Organic matter is uniformly most common accounting for 55–95% of cases [1].
            1. Peanuts and nuts account for 39% (7–58% range).
            2. Organic material has high incidence of associated inflammatory reaction/granulation.
            3. Dried beans, corn, or legumes can swell over time resulting in progressive worsening of obstruction.
        ii. Inorganic material
            1. Plastic objects/toys (5–15%).
            2. School supplies are more common in children over 5 years.
            3. Button batteries are a subset of foreign bodies that represent absolute emergency given potential for rapid, local damage, necrosis, and perforation of tissues. Urgent removal is necessary and specific management strategies and considerations are beyond the scope of this chapter [2].
3. Clinical features [1, 2, 5]
    a. Phases of airway foreign body aspiration
        i. Initial phase—lasting seconds to minutes and characterized by choking, gagging, coughing, respiratory distress, cyanosis, apnea, accessory muscle use, and retractions.
        ii. Asymptomatic phase—occurs after the object lodges and reflexes tire. Can last hours to weeks or longer. Can be relatively asymptomatic or mimic other processes such as asthma, croup, or pneumonia.

    1. During this phase objects can generate inflammation with symptoms responsive to common interventions such as racemic epinephrine, bronchodilators, or corticosteroids resulting in a red herring effect and delaying identification of a foreign body.

   iii. Complication phase—prolonged presence of foreign body leads to chronic obstruction of distal airway, erosion, or infection. Pneumonia, bronchitis, atelectasis, abscess, or fever may all result and further complicate diagnosis and removal of a foreign object.

  b. Location [1, 2]

   i. Laryngeal—less frequent site (2–12%) and most commonly found in children under 1 year of age. This locale and age group represents highest risk of dangerous outcomes.

    1. Partial obstruction symptoms include dysphonia, hoarseness, stridor, cough, sore throat, dysphagia, and sialorrhea.
    2. Complete obstruction symptoms include dysphonia, aphonia, respiratory distress, cyanosis, and respiratory arrest.
    3. Progressive laryngeal edema and/or laryngospasm can transition from partial obstruction to complete if diagnosis is delayed.

   ii. Tracheal—low prevalence (3–12%). Biphasic stridor, croupy cough. Less commonly dysphonia or dysphagia. Described pathognomonic findings:

    1. Audible slap—audible sound during cough or deep inspiration from mobile object impacting tracheal wall.
    2. Palpatory thud—palpation of trachea during the audible slap.
    3. Asthmatoid wheeze—higher pitch and intensity than bronchial asthma and primarily located over the trachea.

   iii. Bronchial—most common (80–90%). Characterized by triad of cough, wheeze, and unilaterally decreased breath sounds. Respiratory distress is relatively uncommon in these patients.

    1. Laterality is more equally distributed in children compared to adults. In adults, right side is more common due to larger diameter and more direct angle of the right mainstem.

4. Diagnosis

  a. History—Witnessed or suspected choking event with coughing or respiratory distress in a previously healthy child [2]. History can be obtained from parents, caregivers, patient, or siblings.

   i. History is prioritized given the potential for physical exam and radiographic evidence to be falsely negative.
   ii. If possible, history should include time of event and type of object. Toys or inorganic objects should be described as best possible or an example

item provided as this can be highly useful in determining best tool for removal.

iii. Pitfalls include unwitnessed events or a child denying event due to fear of punishment [1].

b. Physical Exam [1, 2]

   i. Outward signs include gagging, drooling, dysphonia, cough, stridor (inspiratory vs. biphasic can aid in delineating the location of obstruction), posturing/tripoding, sternal or subcostal retractions, respiratory distress, or anxiety.
   ii. Auscultation.
      1. Laryngeal—inspiratory stridor.
      2. Tracheal—Audible slap or biphasic stridor.
      3. Bronchial—diminished lobar or segmental breath sounds, wheezing.
   iii. Palpation—palpatory thud of mobile tracheal foreign body.
   iv. In the asymptomatic phase, particularly with bronchial foreign bodies, physical exam may be subtle or unrevealing entirely.

c. Radiographic studies [1, 3]

   i. Plain radiographs—first line for any respiratory distress or suspected foreign body.
      1. Ideally would identify a radiopaque object, air trapping, or focal consolidation.
      2. Up to 90% of aspirated foreign bodies are non-radiopaque.
      3. Eighty percent of laryngotracheal foreign body and 30–50% of bronchial foreign body radiographs are read as normal.
   ii. Lateral decubitus films (uncooperative or children <5 years old) or Expiratory films (>5 years old)
      1. Attempt to identify air trapping.
      2. User and patient dependent.
      3. High false-positive rate with questionable benefit [4].
   iii. Fluoroscopy
      1. Dynamic evaluation for mediastinal shift or unilaterally diminished diaphragm excursion secondary to imbalanced airflow in setting of foreign body [3].
      2. Limited utility given availability of CT.
   iv. Computed tomography
      1. Not currently part of the primary foreign body work up due to concerns over radiation exposure and option of bronchoscopy as gold standard for diagnosis.

      2. Can detect non-radiopaque objects, however, mucous plugs can also mimic a foreign body.
      3. Newer scanners are faster, higher resolution, and may not require sedation.

    v. Imaging does aid in diagnosis of complications/sequela associated with retained foreign bodies and remains a component of the evaluation for this reason [1, 3].

  d. Bronchoscopy (rigid or flexible)—gold standard for foreign body diagnosis.

    i. Rigid endoscopy is both diagnostic and provides capacity for intervention, hence it is the preferred modality.

5. Management/Intervention

  a. Preoperative Preparation/Considerations

    i. Suspicion for location of the object dictates urgency of removal. Laryngeal or tracheal objects maintain a risk of complete obstruction and should be dealt with urgently. Objects distal to the carina can be dealt pose less of a mortality risk [2, 6].
    ii. Type of foreign material should also be discerned if possible. Organic materials cans swell with time, oils (particularly from nuts) generate local inflammation/edema/granulation, and sharp objects can pierce the airway [2, 5, 6].

  b. Anesthesia considerations [7]

    i. Clear communication between the surgeon and anesthesia is vital throughout the planning and procedural process. Planning should include methods of induction, ventilation during bronchoscopy, and maintenance of anesthesia.
    ii. Typically, anesthesia is induced with smooth mask ventilation of inhalational agents or spontaneous ventilation with cautious IV induction. For short cases, spontaneous ventilation may be maintained, but for longer or more complex scenarios, neuromuscular suppression may be added with ventilatory support via the rigid bronchoscope. There is no consensus on optimal technique, and this should not be confused with rapid sequence induction/intubation as paralytic agents are not generally given during induction.
    iii. Topical anesthetic is often applied to the glottis and subglottis to prevent reflexes and potential spasm. One percent lidocaine is the most commonly used agent.
    iv. Timing of last meal—added risks of gastric content aspiration. Can consider suctioning of stomach contents after induction but prior to bronchoscopy.
    v. Spontaneous breathing vs. positive pressure ventilation—transitioning from physiologic negative pressure respiration to positive pressure risks

dislodging an unstable foreign body. Maintaining spontaneous ventilation is commonly practiced and forceful positive pressure ventilation is avoided if possible.

c. Equipment [1, 2, 4]

   i. A complete and organized surgical setup is paramount and intervention should not be initiated until equipment is available and in working order. Equipment selection is based on age/size of patient and suspected foreign body involved.

   ii. Laryngoscopes—allow for visualization of the larynx with a working space to allow for retrieval of laryngeal foreign body or insertion of rigid bronchoscope.

      1. Common types include the Parsons laryngoscope (Storz- Fig. 1) and the Tucker side slide laryngoscope (Pilling).
      2. Anesthesia laryngoscopes such as Macintosh, Miller, or Wis blades are also options.

   iii. Bronchoscopes (Fig. 2)

      1. Flexible bronchoscopy is an option for evaluation of the tracheobronchial tree; however, limitations include inability to adequately control an identified foreign body and lack of ability to provide ventilation during the evaluation.
      2. Rigid bronchoscopes combined with Hopkins rod telescopes are the mainstay for tracheobronchial foreign body extraction and evaluation. These come in a wide variety of lengths and diameters and maintain a ventilation port which, when connected to the anesthesia circuit, allows for control of the airway throughout the procedure.

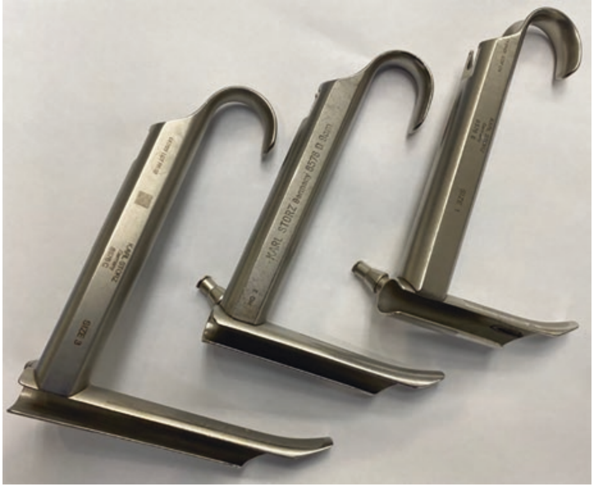

**Fig. 1** Parsons Laryngoscopes (source: Jeremy Mock, MD)

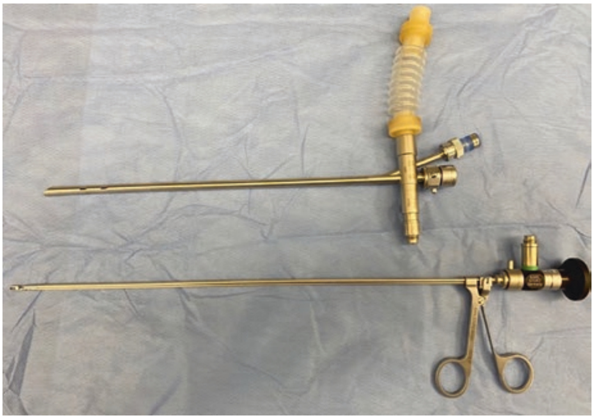

**Fig. 2** Assembled bronchoscope and optical forceps (source: Jeremy Mock, MD)

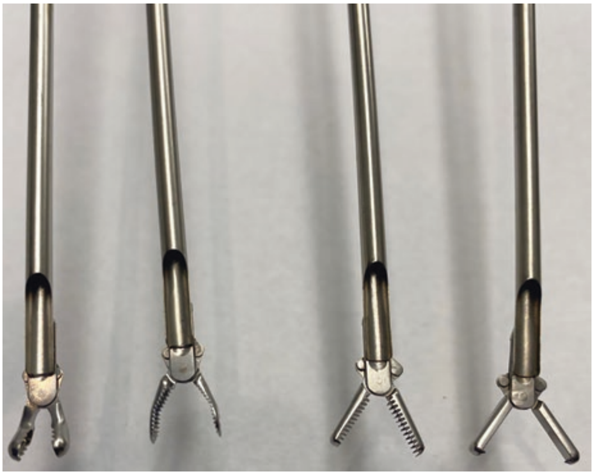

**Fig. 3** Optical forceps graspers from right to left: 1. Cups, 2. Peanut, 3. Straight (alligator), 4. Rat tooth (source: Jeremy Mock, MD)

a. General rule of thumb when selecting the bronchoscope is to choose the largest size that will pass without resistance allowing for maximal visualization and working space.
b. Two bronchoscopes should be lighted and ready in the event that a light fails or forceps become lodged.

iv. Retrieval devices

1. Optical forceps with Hopkins rod telescope should be selected based on the appropriate length for the bronchoscope being used. These should be tested for fit within the bronchoscope prior to the procedure and to ensure the action is working properly.

    a. Many types (Fig. 3) are available including peanut grasping, forward grasping ("alligator"), rat tooth, and cups among many others.

2. Fogarty embolectomy catheter.
    a. In the event of a distally impact foreign body beyond the grasp of the forceps, this small balloon can be inserted past the object, inflated and then withdrawn toward the bronchoscope to present the object so that forceps retrieval is possible.
3. Suction
    a. Having a working suction with flexible catheters passed through the bronchoscopy is imperative.
    b. Although not able to provide sufficient suction to remove most objects, foreign bodies are often shrouded in secretions and suction is necessary to clear the view for extraction.
    c. Organic material can be soft or fragmented and often suction is required to retrieve small pieces or soft material that does not lend itself to grasping with forceps.
4. Special circumstances—despite advances, the rare cases of neonatal and premature infants with foreign bodies are often too small for typical equipment. Creativity with sialendoscopy equipment (scopes and forceps) or cystoscopy tools has been described.

d. Operative Technique [1, 2, 4]
    i. Rigid bronchoscopy is a delicate procedure and, as described, proper setup and preoperative communication is key. Anesthesia should obtain an IV if possible and patient should be rendered under general anesthesia but spontaneously ventilating. Intraoperative dexamethasone should be given.
    ii. The patient should be supine, straight on the operating table, and the table turned toward the surgeon. Shoulder rolls are used at the discretion of the endoscopist but can distort the path of the airway and prevent smooth insertion of the bronchoscope.
    iii. The patient's eyes should be protected and a mouth guard or gauze placed on the alveolar ridge.
    iv. The laryngoscope is inserted on the right side of the mouth, pushing the tongue to the left, and the tip gently inserted into the vallecula.
    v. The laryngoscope is then elevated, lifting the tongue and larynx, taking care to not torque the scope back on the dentition or alveolar ridge.
    vi. If the laryngeal inlet is clear, topical lidocaine (1% or 2%) can be sprayed on the glottis to reduce risk of laryngospasm.
    vii. The bronchoscope can then be inserted gently through the glottis into the upper trachea. The ventilation circuit should be promptly connected, and the laryngoscope removed.
    viii. The tracheobronchial tree is then thoroughly inspected typically beginning with the side uninvolved by the foreign body.

    ix. When a foreign body is identified, its shape, position, characteristics, and mobility should be noted, and proper forceps selected. Suction should be used at this time to remove any associated secretions.
    x. The optical forceps are then utilized through the bronchoscope, the object should be grasped firmly and strategically, and the object gently retracted toward the bronchoscope. If the object is too large to extract through the lumen of the bronchoscope, the scope and forceps should be removed in concert.
    xi. Rarely, if the object is too large to pass the glottis, a tracheotomy is required for removal.
    xii. Following extraction, a second pass should be made to ensure no residual foreign bodies and to assess the airway. Granulation and secretions can be cleared as needed, and bleeding can be controlled with topical vasoconstrictive agents.

e. Postoperative Treatment [4]:

    i. Spontaneously breathing, non-intubated patients are generally awakened and transported to PACU.
    ii. Patients with significant compromise due to edema, narrowing or inflammation associated with the foreign body may require intubation to allow for resolution.
    iii. Antibiotics and steroids are not routinely indicated unless concerning findings are noted during endoscopy [2].
    iv. Postoperative discharge from PACU is feasible in children without preoperative respiratory compromise, protracted work up, and with an uneventful extraction [4].

f. Complications:

    i. Minor complications (12%) include postoperative atelectasis, wheezing, and stridor are usually self-limiting.
    ii. Major complications requiring a week or more of hospitalization were present in 3%.
    iii. Duration of foreign body in the airway is a significant contributor to complications [1].

## Additional Notes

Often labeled “the great mimicker,” pediatric airway foreign bodies represent a broad array of challenges requiring a high index of suspicion and conscientious coordination between multiple pediatric specialists. Despite population-based educational initiatives, preventative measures, and governmental oversite such as the Consumer Product Safety Commission, it remains a relatively frequent pediatric emergency with a potential for fatal consequences. More than 300 children succumb as a result of foreign body aspiration/asphyxiation each year and the incidence of aspiration events, in general, remains steady [5]. Identification and management of airway foreign bodies was a distinct impetus in the creation of aerodigestive endoscopy, and the first described bronchoscopies by Gustav Killian and Algernon Coolidge in the late 1800s were performed in an effort to remove tracheobronchial foreign bodies. Shortly thereafter, Chevelier Jackson developed the lighted bronchoscope as well as adjuvant instruments, removing tens of thousands of objects over the course of his career and inevitably saving thousands of lives. He is appropriately heralded the father of modern endoscopy and the efforts of these pioneers resulted in significant decline in mortality among this population [7].

Any child presenting to the ED, pediatrician, or other providers with complaints of witnessed aspiration event, choking episode, new-onset cough/dysphagia, drooling, dysphonia, or respiratory distress should prompt immediate placement of airway foreign body high on the differential. Recognizing symptoms in each stage of foreign body ingestion and severity of distress dictates urgency of bronchoscopic intervention. The majority of these patients present after the initial “choking” phase, lack signs of complete airway obstruction, and do not represent an acute emergency. As such, thoughtful consideration should involve scheduling endoscopy and removal when trained personnel are available, all instruments are organized and checked, and clear communication between the endoscopist and anesthesia has been conducted [7]. Studies suggest no increased morbidity in stable patients by opting to wait until daytime hours for definitive management [7].

The decision to proceed with rigid bronchoscopy under general anesthesia is one readily undertaken by most seasoned pediatric endoscopists with a common mantra being that negative bronchoscopy is still of high value. Appropriately, alternative options geared at avoiding general anesthesia and the limited risks of rigid endoscopy are emerging. Flexible bronchoscopy under local anesthesia with sedation may be a suitable alternative in a patient without a clear history or exam suggestive of foreign body aspiration [5]; however, identification of a foreign object on flexible scope will likely necessitate transition to rigid bronchoscopy for removal. Additionally, thoracic computed tomography and virtual bronchoscopy are growing in availability and represent an adjuvant to diagnosis for these patients. Early studies suggest high sensitivity and specificity in terms of foreign body identification [7] and would certainly provide confidence in one’s decision to proceed with or defer rigid bronchoscopy. Radiation exposure remains the obvious drawback to this.

Ultimately, prevention is the mainstay to reducing pediatric airway foreign bodies. However, even with the most diligent of caretakers, it is likely impossible to make all such objects inaccessible to children. Continuing education through pediatric offices and well child visits represent some of the best opportunities to educate parents on avoiding the common offending agents such as nuts, popcorn, seeds, and grapes until the child has appropriate dentition and coordination to handle these foods. Encouraging supervised playtime and childproofing is also important.

## Study Questions

1) A 2-year-old, previously healthy male presents to the ED with coughing × 3 days. He is afebrile, behaving normally per mom, and $O_2$ saturations are WNL on room air. Mom denies witnessing any choking events, but he frequently plays with Legos with his older brother. Physical exam is suggestive of diminished lung sounds on the right lower lobe. Chest X-ray, including lateral decubitus films are read as normal. The ED felt that albuterol helps the wheezing they heard on exam. What is the next step in management?

   a. CT scan of the chest
   b. Flexible bronchoscopy
   c. Rigid bronchoscopy in the OR
   d. Antibiotics and follow up

Answer to Question 1:

c. Rigid Bronchoscopy—index of suspicion should be high in this age group, particularly with a previously healthy patient. Although no witnessed event occurred, his history with small objects and focal exam findings are concerning. Negative imaging does not preclude the need for bronchoscopy and rigid exam allows for both diagnosis and intervention without subjecting him to radiation from a CT scan.

2) A 3-year-old female presents to the ED at 11 pm with witnessed choking event when eating dinner at 8 pm consisting of carrots and almonds. Mom performed back blows and she coughed a piece of carrot. She has no symptoms since that time, has diffuse wheezes in all lung fields on auscultation, and X-ray suggests bilateral consolidations. Lateral decubitus films show hyperinflation on the right. How do you proceed?

   a. Take to the OR stat despite lack of NPO status. This is clearly an aspiration and needs to be dealt with.
   b. Reassure mom and discharge explaining that the wheezing is reactionary to the event.
   c. Admit overnight for monitoring and take to the OR, electively, the next day for rigid bronchoscopy.
   d. Consult Pulmonology for asthma exacerbation.

Answer to Question 2:

c. OR the next day. Bronchial foreign bodies do not require urgent surgery if the proper teams and equipment are not readily available. It is much safer to wait until the OR is fully staffed and all necessary team members are in place. This also allows for her to be properly NPO for the procedure. Although she may have coughed out the foreign body, exam findings suggest otherwise and removing organic material sooner results in less inflammation and risk of complications. A negative bronchoscopy in skilled hands is also a low-risk intervention.

## References

1. Bluestone CD, Simons JP, Healy GB. Chapter 96: Foreign bodies of the larynx, trachea, and bronchi, pp 1609–1625. Bluestone and stool's pediatric otolaryngology, 2014.
2. Rodríguez H, Passali GC, Gregori D, Chinski A, Tiscornia C, Botto H, Nieto M, Zanetta A, Passali D, Cuestas G. Management of foreign bodies in the airway and oesophagus. Int J Pediatr Otorhinolaryngol. 2012;76(Suppl 1):S84–91. https://doi.org/10.1016/j.ijporl.2012.02.010.
3. Hegde SV, PKT H, Lee EY. Tracheobronchial foreign bodies in children: imaging assessment. Semin Ultrasound CT MRI. 2015;36(1):8–20. https://doi.org/10.1053/j.sult.2014.10.001.
4. Andreoli SM, Kofmehl E, Sobol SE. Is inpatient admission necessary following removal of airway foreign bodies? Int J Pediatr Otorhinolaryngol. 2015;79(9):1436–8. https://doi.org/10.1016/j.ijporl.2015.06.020.
5. Foltran F, Ballali S, Passali FM, Kern E, Morra B, Passali GC, Berchialla P, Lauriello M, Gregori D. Foreign bodies in the airways: a meta-analysis of published papers. Int J Pediatr Otorhinolaryngol. 2012;76(Suppl 1):S12–9. https://doi.org/10.1016/j.ijporl.2012.02.004.
6. Gang W, Zhengxia P, Hongbo L, Yonggang L, Jiangtao D, Shengde W, Chun W. Diagnosis and treatment of tracheobronchial foreign bodies in 1024 children. J Pediatr Surg. 2012;47(11):2004–10. https://doi.org/10.1016/j.jpedsurg.2012.07.036.
7. Fidkowski CW, Zheng H, Firth PG. The anesthetic considerations of tracheobronchial foreign bodies in children. Anesth Analg. 2010;111(4):1016–25. https://doi.org/10.1213/ANE.0b013e3181ef3e9c.

# Congential Cervical Lesions

**Randy Lesh, Curtis Gehman, and Aileen P. Wertz**

**Key Points**

1. Providers should maintain a high index of suspicion for benign congenital neck masses when children present with neck abscesses or other neck masses.
2. Determining the specific congenital neck mass is determined primarily by location, appearance, and palpation of the mass on physical exam.
3. Surgical excision is the standard of care for most common congenital neck masses.
4. In addition to branchial anomalies and thyroglossal duct cysts, other benign congenital neck masses that should be considered for the differential diagnosis include dermoid cyst, pseudotumor of infancy, thymic cyst, cartilage rest, and the continuum of venolymphatic malformations. Malignancies and infectious etiologies must also be considered in the differential diagnosis for pediatric neck masses.

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## Thyroglossal Duct Cyst

1. Epidemiology
    a. Thyroglossal duct cysts (TGDC) are the most common cause of midline anterior neck masses in children.
    b. TGDCs have a bimodal age distribution occurring most commonly in the first and fifth decades of life with an incidence of 2.2/100,000 [1].
2. Pathophysiology
    a. Thyroid anlage migrates from the base of the tongue at the foramen cecum either anteriorly, posteriorly, or through the hyoid bone to the midline neck, where it reaches its destination at the end of the 7th week of gestation. As it descends it leaves behind an epithelial-lined tract remnant.
    b. Usually, this thyroglossal tract atrophies and disappears between 5- and 10-weeks' gestation. If the tract does not disappear, thyroid remnants may remain and form a cyst or fistula at any site along the route [2].
3. Clinical Features
    a. History:
        i. Painless, midline neck mass which may fluctuate in size.
        ii. Infected cysts may be erythematous, tender, and draining.
        iii. Rarely, can present with hypothyroidism if no thyroid gland is present.
    b. Physical Exam:
        i. Soft to firm, mobile, non-tender mass in the midline of the neck.
        ii. Most are located at the level of the hyoid or infrahyoid [3].
        iii. Usually 2–4 cm [4].
        iv. Elevates with protrusion of the tongue.
4. Diagnostics (Fig. 1)
    a. Ultrasound is the imaging of choice. It is done to help determine presence of thyroid gland and distinguish between ectopic thyroid tissue, dermoid cyst, branchial cleft cysts, enlarged lymph node, and hemangioma [3, 5].
    b. CT and MRI are usually unnecessary for pediatric patients but can be used as a supplement to ultrasound [3].
    c. Radioisotope scan of the thyroid gland: Indicated if there are signs and symptoms of hypothyroidism or absence of thyroid gland on ultrasound. If this is the child's only functioning thyroid tissue, removal will leave the patient hypothyroid. This would be a contraindication for excision [5, 6].
    d. Thyroid function tests can be done if there are signs and symptoms of hypothyroidism or if the thyroid gland is not seen on ultrasound [4, 6].
    e. Fine needle aspiration (FNA) plays a limited role and is rarely used to confirm diagnosis.

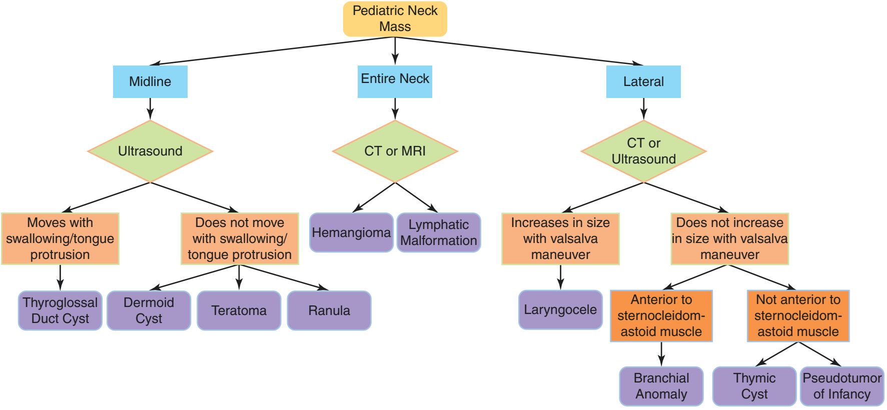
Pediatric Neck Mass
Midline
Entire Neck
Lateral
Ultrasound
CT or MRI
CT or Ultrasound
Moves with swallowing/tongue protrusion
Does not move with swallowing/ tongue protrusion
Hemangioma
Lymphatic Malformation
Increases in size with valsalva maneuver
Does not increase in size with valsalva maneuver
Thyroglossal Duct Cyst
Dermoid Cyst
Teratoma
Ranula
Laryngocele
Anterior to sternocleidom- astoid muscle
Not anterior to sternocleidom- astoid muscle
Branchial Anomaly
Thymic Cyst
Pseudotumor of Infancy

**Fig. 1** Algorithm for working up pediatric neck masses (Source: Randy Lesh, Curtis Gehman, Aileen Wertz)

5. Treatment:
    a. TGDC presenting infected should first be treated with antibiotics. Antibiotic coverage is the same as for upper respiratory infections and therefore Augmentin or its IV counterpart are often first line.
    b. Incision and drainage (I&D) may be required if the TGDC has become large due to infection resulting in mass effect symptoms, or if antibiotics are not resolving the infection.
    c. Therapeutic FNA is an intermediary option often preferred over I&D because it results in less difficulty with future complete excision than I&D [7].
    d. Indications for surgery:
        i. High rate of infection if not excised.
        ii. 1–2% of thyroglossal duct cysts can become malignant.
        iii. Cosmetic appearance [7].
    e. Operative approach: Sistrunk procedure.
        i. General anesthesia is given, and an endotracheal tube is inserted.
        ii. The patient is placed in a supine position with the neck hyperextended with the midline neck exposed the area is prepped in the sterile fashion.
        iii. A transverse incision over the cyst and/or in a relaxed skin tension line is made; if a draining sinus is present, an elliptical incision is made to excise the sinus.
        iv. The strap muscles are exposed and divided vertically in the midline.
        v. The cyst and duct remnant are identified and dissected to the hyoid bone where the sternohyoid and thyrohyoid muscle attachments are divided enough to see the path of the thyroglossal duct. About 1 cm of the central part of the hyoid, through which the duct remnant passes, is removed. The mylohyoid and geniohyoid muscle attachments are also divided.

        vi. Dissection may need to be carried out superiorly to the foramen cecum at the base of the tongue if duct is visualized superior to the excised hyoid bone and can be traced to the foramen cecum. The end of the duct remnant is suture ligated [8].
        vii. The hyoid is left in discontinuity.
        viii. The wound is closed in a multilayered fashion with absorbable suture.
        ix. The specimen should be sent to pathology for evaluation.
        x. Prophylactic antibiotic use is controversial but should not be needed unless acute infection present.

6. Outcomes
    a. The most common complication is incomplete removal of the TGDC resulting in recurrence. Recurrence rate is estimated between 5 and 33% with significantly fewer recurrences when Sistrunk procedure is performed [7].

## Branchial Anomalies

1. Epidemiology: Account for about 17% of pediatric neck masses.
2. Pathophysiology
    a. Incomplete obliteration and resorption of a branchial (pharyngeal) apparatus (BA) lead to the formation of a branchial cleft cyst, sinus, and/or fistula.
    b. Paired branchial arches and associated clefts and pouches begin to develop during the second week of gestation and are well-defined by the fourth week. They form the structures of the head and neck as given in Table 1.
    c. Types of branchial anomalies
        i. Branchial cyst—No internal or external opening
        ii. Branchial sinus—Open to the skin but does not communicate with the pharynx
        iii. Branchial fistula—Communication between the aerodigestive tract and the skin. Table 2 describes these in greater detail.
3. Clinical Features
    a. History:
        i. Present at birth but typically noted in late childhood or early adulthood following infection (frequently post-upper respiratory tract infection).
        ii. May present with drainage and pain if infected.
    b. Physical Exam:
        i. Palpable cyst in the lateral neck, most commonly found along the anterior border of the sternocleidomastoid muscle (2nd BA) (Fig. 2).
        ii. Tender on palpation if currently infected and non-tender if uninfected.

**Table 1** Structures derived from paired branchial arches

| Arch | Pouch derivative | Nerve | Muscle(s) | Skeletal structure(s) | Artery |
|---|---|---|---|---|---|
| First (mandibular) | Eustachian tube, middle-ear cavity, mastoid air cells | Trigeminal (V) | Muscles of mastication, mylohyoid, anterior digastric, tensor tympani, tensor velum palati | Malleus, incus, portion of mandible | Maxillary |
| Second (hyoid) | Palatine tonsils, supratonsillar fossa | Facial (VIII) | Muscles of facial expression, stapedius, stylohyoid, posterior digastric | Stapes, styloid, lesser cornu of hyoid, upper body of hyoid | Stapedial |
| Third | Inferior parathyroid glands | Glossopharyngeal (IX) | Stylopharyngeus | Greater cornu of hyoid, lower body of hyoid | Common and internal carotid |
| Fourth | Superior parathyroid glands, thymus | Superior laryngeal (X) | Pharyngeal constrictors, cricothyroid | Laryngeal cartilages | Subclavian on right, arch of aorta on left |
| Sixth | – | Recurrent laryngeal (X) | Intrinsic laryngeal muscles | Laryngeal cartilages | Pulmonary artery on right, ductus arteriosus on left |

**Table 2** Characteristics and prevalence of branchial anomaly types

| Branchial anomaly | Location | Fistula Tract | Prevalence |
|---|---|---|---|
| First (Type I) | External auditory canal | Pass close to facial nerve | ~20% |
| First (Type II) | Below angle of mandible | Through parotid gland, lateral or medial to facial nerve, and end within or inferior to external auditory canal | |
| Second | Anterior to sternocleidomastoid muscle (high in neck) | Deep to second arch structures, superficial to third arch structures, and end in tonsillar fossa | ~75% |
| Third | Anterior to sternocleidomastoid muscle (low in neck) | Deep to third arch structures, superficial to fourth arch structures, and enter pharynx at thyrohyoid membrane or piriform sinus | ~5% |
| Fourth | Lower neck/adjacent to thyroid gland | Open to piriform sinus and course inferior to superior laryngeal nerve | |

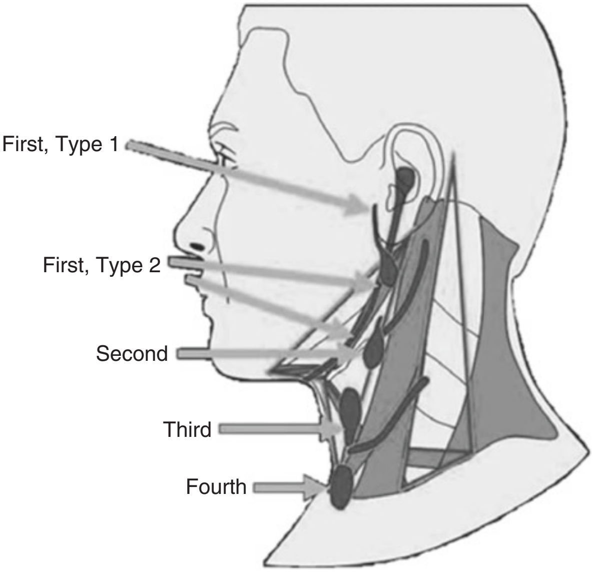

**Fig. 2** Location of branchial cleft remnants. Branchial cleft cysts and sinuses are classified based upon the level of branchial cleft remnant from which they arise (Source: Adapted from Dr. James Smirniotopoulos, with permission, via http://rad.usuhs.edu/medpix/cow_image.html?pt_id=13954&imid=56175&quiz=no&week=590#top. Downloaded Dec 27, 2014)

        iii. Possible opening in skin.
        iv. Expression of mucus/mucopurulent fluid upon palpation if open to skin.

4. Imaging:
    a. Ultrasonography is often obtained initially, especially in the context of acute infection and uncertain diagnosis. Starting with CT or MRI is reasonable for lateral neck masses unless pseudotumor of infancy is the most likely diagnosis.
    b. CT or MRI neck with contrast is often obtained prior to considering surgical excision to most thoroughly identify the extent of BA remnant and its relationship to other head and neck structures, especially if there is a sinus opening in skin suggesting more extensive fistula.
    c. MRI will provide the best soft tissue detail of any fistula present, but still may not demonstrate the full extent of a fistula; therefore, the surgeon must be prepared for a fistula excision even if imaging does not demonstrate one.

5. Treatment:
    a. See above discussion under treatment of TGDC for recommendations for managing infection and rationale for elective surgical excision.
    b. Elective surgical excision.
        i. The exact approach depends on whether there is a fistula tract or just cyst, and from which branchial arch the mass arises. Below are general principles.
        ii. Injection of the sinus with methylene blue dye and/or insertion of a lacrimal probe aids in identifying and dissecting fistulous tract.

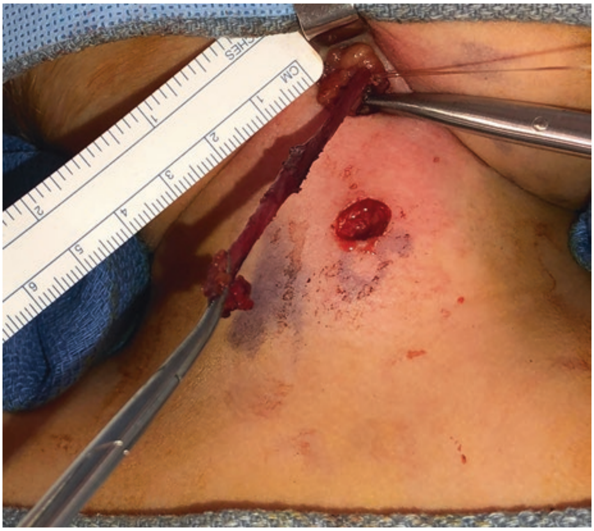

**Fig. 3** Surgical excision of a second branchial cleft cyst for using the “stepladder” technique (Source: Marc Lessin)

   iii. If sinus/fistula tract is present, then elliptical skin incision to excise sinus is required. If there is only a cyst, then a horizontal skin incision overlying the cyst or in a nearby relaxed skin tension line is best.
   iv. Second incision superior to the first in a stepladder fashion may be necessary to expose tract superiorly (Fig. 3).
   v. Fistula tracts require extensive dissection and careful protection of nearby structures. See Table 2 for structures that the surgeon can anticipate encountering during dissection.
      1. First brachial fistulas run near facial nerve branches.
      2. Second branchial fistulas are in proximity to the hypoglossal nerve, hypopharyngeal nerves, and the bifurcation of the carotid artery.
      3. Third and Fourth branchial fistulas require inspection of the piriform sinus before surgical exploration, with possible removal of the posterior portion of the thyroid cartilage to gain exposure.
   vi. Excision of entire cyst/sinus and fistula tract should be performed to prevent recurrence.

c. Endoscopic cauterization of the piriform sinus opening rather than complete fistula tract excision is an option for third and fourth arch anomalies.
   i. The skin sinus can also be cauterized.
   ii. Bipolar cautery is typically used.
   iii. This technique decreases surgical risk and has been demonstrated to resolve draining fistula tract in a couple case series.
   iv. There is a risk of recurrent laryngeal nerve injury when bipolar is used in the piriform sinus. Recurrent laryngeal nerve monitoring is recommended and bipolar should be on the lowest effective setting.

6. Complications and Prognosis
    a. Potential damage to nearby nerves and arteries of the head and neck.
    b. Recurrence is uncommon if complete excision is performed and in patient without infection or previous neck surgery, both of which make complete excision more difficult.

## Additional Notes

In this chapter, we discussed workup and treatment of 2 of the most common congenital cervical lesions, thyroglossal duct cysts and branchial anomalies. There are many other causes of cervical lesions in children including infections, malignancies, and other, less common, congenital anomalies.

It is important to consider a congenital cyst when children present with a neck abscess that appears to be unilocular, or in a classic location for one of these congenital cysts. This helps set expectations for possible recurrence and need for further surgery. It also prompts consideration for needle aspiration or antibiotics alone, when safe and appropriate. If incision and drainage can be avoided, then definitive excision is easier. Management of infected congenital cervical cysts is more conservative than it was in the past with the goal of keeping definitive excision incisions as small as possible and morbidity as minimal as possible.

## Study Questions

1) What is the most common anterior neck mass in children?
    A.) Branchial cleft cyst
    B.) Dermoid cyst
    C.) Thyroglossal duct cyst
    D.) Plunging ranula

Answer to Question 1:

(C) Thyroglossal duct cyst is the most common anterior neck mass in children. Branchial cleft cysts usually present as a lateral neck mass. While dermoid cysts are also an anterior neck mass, they do not elevate with tongue protrusion and are not as common as TGDC. Plunging ranulas are pseudocysts of the floor of mouth that may present as an anterior neck mass but are less common.

2) What is the preferred treatment with the lowest rate of reoccurrence for uncomplicated thyroglossal duct cysts?
    A.) Incision and drainage
    B.) Sistrunk procedure
    C.) Antibiotics alone
    D.) Central neck dissection

Answer to Question 2:

(B) The Sistrunk procedure, which involves removal of the tract and middle portion of the hyoid bone, is the preferred surgical treatment of thyroglossal duct cysts. Incision and drainage can be used for infected cysts but do not remove the cyst itself. Treatment with antibiotics alone may resolve an acute infection of a cyst, but chronic infections are common. Central neck dissection is usually not required in the initial surgical procedure for uncomplicated TGDC but may be used in select cases (7).

3) An 11-year-old boy presents to the office with a painful mass on the right side of his neck for 2 weeks. There is an opening in the skin overlying the lump with mucopurulent drainage. He denies any recent trauma to the neck. The mass does not move with tongue protrusion. Which of the following is most likely to be elicited upon taking further history?

   A.) Presence of this neck lump since birth
   B.) History of similar neck lumps
   C.) Family history of malignancy
   D.) Dysphagia
   E.) Recent upper respiratory tract infection

Answer to Question 3:

(E) Recent upper respiratory tract infection. The presence of a painful lateral neck mass with mucopurulent drainage and no movement with tongue protrusion is most consistent with an infected branchial cleft cyst. Branchial cleft cysts are present at birth but most commonly present after becoming infected. Branchial cleft cysts most commonly become infected following an upper respiratory tract infection.

4) A 7-year-old girl underwent elective surgical excision of a benign congenital neck mass 1 week ago. She presents for follow-up evaluation. Examination reveals tongue deviation to the left on protrusion. This patient most likely had which of the following congenital neck masses:

   A.) Thyroglossal duct cyst
   B.) Right second branchial cleft cyst
   C.) Left second branchial cleft cyst
   D.) Right first branchial cleft cyst
   E.) Left first branchial cleft cyst

Answer to Question 4:

(C) Left second branchial cleft cyst. This patient is likely suffering from left hypoglossal nerve injury following surgical excision of a second branchial cleft cyst. Second branchial cleft cysts commonly course near the hypoglossal nerve, creating potential for nerve damage during surgery. The presentation of tongue deviation to the left suggests right hypoglossal nerve damage due to preserved function of the right hypoglossal nerve.

## Bibliography

1. Thompson LDR. Thyroglossal duct cyst. Ear Nose Throat J. 2017;96(2):54–5.
2. Allard RH. The thyroglossal cyst. Head Neck Surg. 1982;5(2):134–46.
3. Lim-Dunham JE, Feinstein KA, Yousefzadeh DK, Ben-Ami T. Sonographic demonstration of a normal thyroid gland excludes ectopic thyroid in patients with thyroglossal duct cyst. AJR Am J Roentgenol. 1995;164(6):1489–91.
4. Kathuria B. Thyroglossal duct cyst & sistrunk, a case series; Personal experience and literature review. J Otolaryngol ENT. 2015;3(3)
5. Brewis C, Mahadevan M, Bailey CM, Drake DP. Investigation and treatment of thyroglossal cysts in children. J R Soc Med. 2000;93(1):18–21.
6. Tunkel DE, Domenech EE. Radioisotope scanning of the thyroid gland prior to thyroglossal duct cyst excision. Arch Otolaryngol Head Neck Surg. 1998;124(5):597–9.
7. Chou J, Walters A, Hage R, et al. Thyroglossal duct cysts: anatomy, embryology, and treatment. Surg Radiol Anat. 2013;35(10):875–81.
8. Righini CA, Hitter A, Reyt E, Atallah I. Thyroglossal duct surgery. Sistrunk procedure. Eur Ann Otorhinolaryngol Head Neck Dis. 2016;133(2):133–6.
9. Flint PW, Cummings CW. Congenital and inflammatory neck masses in children. In: Cummings otolaryngology: head and neck surgery. 7th ed. Philadelphia, PA: Elsevier; 2021. p. 2989–3001.
10. Johnson JT, Rosen CA, Bailey BJ. Congenital neck masses and cysts. In: Bailey's head and neck surgery--otolaryngology, vol. I. 4th ed. Philadelphia, PA: Wolters Kluwer Health/ Lippincott Williams & Wilkins; 2014.
11. Coste AH, Lofgren DH, Shermetaro C. Branchial cleft cyst. 2021 Jan 8. In: StatPearls [Internet]. Treasure Island, FL: StatPearls Publishing; 2021. Accessed 24 OCT 2021.

# Renal Duplication Anomalies

**Jasmine I. Kashkoush and Robert Steckler**

**Key Points**

1) In a duplicated system, the ectopic ureter is usually the upper pole ureter.
2) The Weigert-Meyer rule describes the relationship of the lower pole moiety kidney that is laterally displaced and is prone to VUR. The upper pole moiety is medially displaced and is prone to obstruction.
3) Majority of renal duplication anomalies are found prenatally on ultrasound imaging. Hydronephrosis is the most common finding.

Renal Duplication Anomalies pose a challenge for medical providers due to their relatively silent nature. Many are detected prenatally during routine maternal-fetal US. However, patients may develop symptoms of infection or obstruction after birth.

1. Pathophysiology
    a. Epidemiology
        i. Very common
        ii. Estimate 1/50 population has a duplex collecting system
    b. Definition
        i. Renal Duplication
            1. Complete—Two separate ureteral orifices for drainage into bladder.

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            2. Incomplete—Two ureters join together proximal to the bladder, while one ureter enters the bladder.

        ii. Ureterocele

            1. Cystic dilation of distal intravesical ureter
            2. Can be associated with a single system: described as orthotopic. Non-obstructive.
            3. Duplex system

                i. Ectopic ureterocele
                ii. Cecoureterocele: The ureterocele extends into the bladder neck and possibly the urethra.

            4. Essential to assess which moieties are in jeopardy

                i. Ipsilateral upper pole only.
                ii. Ipsilateral upper and lower poles.
                iii. Ipsilateral and contralateral upper tracks—Secondary to ectopic ureterocele or cecoureterocele obstructing the bladder neck/urethra.

        iii. Ectopic ureter

            1. Inserts off of its orthotopic position on the trigone.
            2. Lateral ectopia within bladder results in a short submucosal tunnel of the ureter and predisposes to vesicoureteral reflux (VUR).
            3. Caudal ectopia may result in varying degrees of obstruction and associated renal dysplasia.
            4. In girls, the ectopic ureter may drain outside of the urinary tract altogether, into the vagina or perineum, potentially producing global urinary incontinence.
            5. In boys, the ectopic ureter cannot drain beyond the external sphincter. May drain into Wolffian structures.

    c. Weigert-Meyer Law

        i. Describes the relationship between duplication and ectopia.
        ii. The ureteral orifice from lower pole moiety kidney is laterally displaced and is prone to VUR—can also have concomitant UPJO.
        iii. The ureteral orifice from upper pole moiety kidney is medially displaced and is prone to obstruction.

2. Clinical features

    a. May present with infection.
    b. Urinary incontinence may be caused by an ectopic ureter that bypasses the urethral sphincter in a girl but not in a boy. Males have rare problems with epididymitis.
    c. Rare, but ureterocele may prolapse and be visualized between the labia.

3. Diagnosis
    a. Majority were detected via prenatal ultrasound imaging. Hydronephrosis is the most common finding.
    b. Evaluate with renal/bladder ultrasound and VCUG.
    c. MAG3 Lasix (furosemide) renal scan if concerned about obstruction.
4. Treatment
    a. Many options for repair of this diverse spectrum of anomalies.
        i. Goals: Preservation of renal function, elimination of infection, obstruction, and reflux, and maintaining urinary continence.
    b. Management of the neonate with Duplication Anomaly.
        i. Common prenatal detection.
        ii. Use antibiotic prophylaxis if significant VUR or hydronephrosis.
    c. Early surgical intervention usually focuses on relief of obstruction.
        i. Primarily from ectopic ureterocele.
        ii. Usually approached endoscopically.
    d. Transurethral incision of an ectopic ureterocele.
        i. Used to relieve obstruction.
            1. Usually of upper pole ureter and kidney.
            2. Occasionally of the bladder outlet.
        ii. Can use Bugbee electrode (Medpro Medical, Veenendaal, The Netherlands) or holmium laser.
        iii. Can create iatrogenic VUR.
        iv. In cases of severe VUR other moieties can be more aggressive.
        v. Must be careful in cases of cecoureterocele not to create a flap that can obstruct the flow of urine and create bladder outlet obstruction.
    e. Surgical Management of Child.
        i. Primarily associated with correction of hydronephrosis and stasis of urine responsible for RUTIs and possible renal scarring.
        ii. Must have a high index of suspicion for the ectopic ureter in girls that could result in continuous incontinence once potty-trained.
            1. In the past presented typically around ages 5–9 years old.
            2. MRI is helpful in delineating this complex anatomy: must inform pediatric radiologist of suspicions so study can be tailored.
        iii. Preserve renal parenchyma if functioning well, however, dysplasia is common and most often involves excision of the poorly functioning segment.

    iv. Most commonly this is the upper pole moiety but occasionally the lower pole if associated with severe VUR.
    v. Do not address bladder and VUR initially as in a total reconstruction.

f. Total Reconstruction.

    i. Most often upper pole (UP) heminephrectomy with ureterocele excision and reimplantation of lower pole ureter is performed.
    ii. Can be approached open via flank extraperitoneal incision or laparoscopically: use of robotic assistance is helpful.
    iii. Avoid damage to the lower pole (LP).
    iv. Can evaluate postoperatively with a Doppler sonogram to rule out urinoma and show blood flow to the lower pole.
    v. Pre-stent normal ureter to aid in identification and preservation.
    vi. Great care mobilizing the UP megaureter under the renal vessels.

        1. Undue tension on these small vessels can result in spasm or thrombosis.
        2. Less likely with laparoscopic approach.

    vii. Usually, small branch to UP segment, which once divided shows clear demarcation for excision.
    viii. Must be aware that LP segment can be injured if not careful and result in bleeding and/or entry collecting system.
    ix. UP ureter is excised down to pelvis where the two ureters tend to share a common wall.
    x. Stent is removed at end of case.
    xi. Estimated that 66% of duplication anomalies can be handled with renal approach alone.
    xii. Additional third will need bladder reconstruction, primarily for severe VUR.

g. Bladder Reconstruction in Duplication Anomalies.

    i. Must address severe VUR and or ureteroceles.
    ii. The ureterocele is excised.
    iii. Ipsilateral and even the contralateral ureters may need to be reimplanted.
    iv. Can result in a large defect in the base of the bladder.

        1. This must be closed.
        2. Take care to avoid injury to underlying vagina.

    v. Use of JJ-stenting is helpful.
    vi. Usually, can manage with Foley catheterization post-op.
    vii. Close follow-up for UVJ obstruction and/or persistent VUR.

## Additional Notes

Given the broad range of pathology with renal duplication anomalies, prior to any surgical intervention, it is important to gain as much information regarding the altered anatomy and its impact. The goal of surgery is preservation of renal function, elimination of infection, obstruction, and reflux, and maintaining urinary continence.

With most of the pathology we see, early intervention with prophylactic antibiotics is generally warranted to minimize the risk of infection.

## Study Questions

1) Question 1: True or False: An ectopic ureter in a male typically presents with urinary incontinence.

   a. True
   b. False

Answer to Question 1: (b.) False. In a male, the ectopic ureter never inserts past the external urinary sphincter and thus, they do not present with urinary incontinence. If females, the ectopic ureter may drain outside of the urinary system and thus, they may present with urinary incontinence.

2) Question 2: The Weigert-Meyer rule describes the relationship of the lower and upper pole moiety kidney. The lower pole ureter is usually:

   a. Laterally displaced
   b. Medially displaced

Answer to Question 2: (a.) Laterally displaced. The Weigert-Meyer rule describes the relationship of the lower pole moiety kidney that is laterally displaced and is prone to VUR. The upper pole moiety is medially displaced and is prone to obstruction.

3) True or false: In a duplicated system, the ectopic ureter is usually the upper pole ureter.

   a. True
   b. False

Answer to Question 3: (a.) True. The Weigert-Meyer rule describes the relationship of the lower pole moiety kidney that is laterally displaced and is prone to VUR. The upper pole moiety is medially displaced and is prone to obstruction.

## Further Reading

Kalfa N, author, Weiss DA, editor. Congenital anomalies of the kidney and urinary tract. AUA University. American Urological Association. Linthicum, MD. 2021. https://university.auanet.org/core/pediatric/congenital-anomalies-of-the-kidney-and-urinary-tract/index.cfm, Accessed 6 NOV 2021.

Partin AW, Dmochowski RR, Kavoussi LR, Peters CA, Wein AJ. Campbell Walsh Wein urology. 12th ed. Amsterdam, NL: Elsevier; 2020.

Smith JA, Howards SS, Preminger GM, Dmochowski RR. Hinman's Atlas of urologic surgery revised reprint. 4th ed. Amsterdam, NL: Elsevier; 2019.

# Cryptorchidism

**Jasmine I. Kashkoush and Robert Steckler**

**Key Points**

1) Ensure a thorough physical examination is completed with the child relaxed and, in several positions, to confirm incomplete descent of the testis.
2) The gold standard for diagnosis is a physical examination. A scrotal ultrasound is not needed for the diagnosis or management.
3) If spontaneous testicular descent is not achieved by 6 months, surgical treatment is warranted.

## Cryptorchidism

Cryptorchidism or undescended testis (UDT) is the absence of one or both testes in a normal scrotal position. This is one of the most common congenital anomalies, occurring in 3% of full-term and up to 45% of preterm newborn males.

1. Pathophysiology
    a. Incidence
        i. Up to 45% incidence in preterm and small for gestational age males.
        ii. Approximately 3% incidence in full-term infants.
        iii. At 6 months old, incidence is approximately 1%.

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b. Risk factors
    i. Prematurity and/or low birth weight for gestational age.
    ii. Familial factors (maternal environment).
    iii. Maternal diabetes.
    iv. Maternal smoking.
c. Virtually all testicles that descend spontaneously will do so within 3 months, correcting for any prematurity. Those that descend do not require operative treatment.
d. UDT is usually unilateral.
e. Acquired UDT (pseudocryptorchidism) occurs after prior inguinal surgery when the testicle has become entrapped above the scrotum.
f. Ascended testicles are testicles that were previously fully descended, most commonly representing ascent of a previously retractile testicle.
g. Anatomic location: May be located anywhere along the line of normal descent from the lower pole of the kidney in the retroperitoneum, the inguinal canal and scrotum or in an ectopic position.
    i. Intra-abdominal (nonpalpable UTD)
    ii. Peeping: At the level of the internal ring
    iii. Canalicular: Within the inguinal canal (most common)
    iv. High scrotal or retractile testis
    v. Ectopic locations
        1. Anterior to the rectus muscle (SIP)
        2. Prepubic
        3. Femoral
        4. Peri-penile
        5. Perineal
        6. Contralateral scrotum
h. Pathogenesis
    i. Unilateral UDT
        1. Majority are unilateral (60–70%), generally more common on the right.
        2. Usually developmentally normal testicles.
        3. Testicles that remain undescended beyond a year of age begin to lose germ cells. Paternity rates for men with unilateral UDT are statistically the same as men born with two descended testicles.
    ii. Bilateral UDT
        1. Increased incidence of subfertility.
        2. May be associated with an anatomic anomaly that is responsible for the UDT.

- i. Associated Conditions:
    - i. Defects of androgen synthesis or receptors
    - ii. Disorders of sexual differentiation
    - iii. Klinefelter's syndrome
    - iv. Down's syndrome
    - v. Prune-belly (Eagle-Barrett Syndrome)
    - vi. Omphalocele/gastroschisis
    - vii. Bladder exstrophy
    - viii. Hypospadias

2. Diagnosis
    - a. Infants and boys should be examined supine, and if necessary cross-legged/catcher's position. Abducting the thighs inhibits the cremasteric reflex which can facilitate drawing the testicle into the scrotum.
    - b. Techniques to help with a proper examination:
        - i. Distracting patients
        - ii. Warm examination room
        - iii. Having warm hands
        - iv. Using liquid soap or lubricant
        - v. Repeat exams
    - c. Retractile testes are the most common diagnosis in differential.
        - i. Retractile testicles can be manipulated into the scrotum and will stay down, whereas an UDT cannot be drawn into the scrotum and/or will fail to stay down when released.
        - ii. Ipsilateral hemiscrotum is normally well developed.
        - iii. Is normal with respect to spermatogenesis.
        - iv. Has no increased risk of malignancy.
        - v. If unclear (especially with hyperactive or anxious children), repeat exam in another setting.
        - vi. Provide parents with reassurance.
    - d. Classify palpable vs. non-palpable UDT
        - i. Guides surgical approach
    - e. Palpable UDT
        - i. Majority UDTs (75–80%).
        - ii. Most are low or present in canal. Some can be "peeping" in and out of internal ring.
        - iii. Almost all can be approached surgically in standard fashion with inguinal orchiopexy.

f. Non-palpable UDTs

   i. Approx. 20–25% of all UDTs.
   ii. May be difficult to palpate in obese boys. Gold standard: exam under anesthesia.
   iii. No indication for routine imaging.
   iv. Possible findings at surgery:

      1. Abdominal or peeping location 25–50%.
      2. Complete atrophy or "vanishing" 15–40% (secondary to perinatal torsion). Diagnose by documenting blind ending spermatic vessels at exploration.
      3. Extra-abdominal location 10–30%.

3. Treatment

   Bilateral impalpable testicles—draw blood for Anti-Mullerian Hormone. If measurable, surgical exploration is necessary. If AMH is not detected, surgery is unnecessary as the patient is anorchid (Fig. 1).

   a. Orchiopexy is indicated to facilitate testicular examination for tumor surveillance, optimize spermatogenesis, provide cosmetic benefit, and repair associated hernia if present.
   b. Infants should be followed for spontaneous descent until 6 months of age.
   c. There is no indication for observation after this time point.
   d. There is no current indication for hormonal treatment to stimulate testicular descent.
   e. When a palpable UDT presents in pubertal and postpubertal boys, testicular biopsy should be considered because of the 3% incidence of carcinoma in situ. Fifty percent of these patients may go on to develop testicular cancer. Orchiectomy may be considered as well. Can place testicular prosthesis in the same setting if desired.

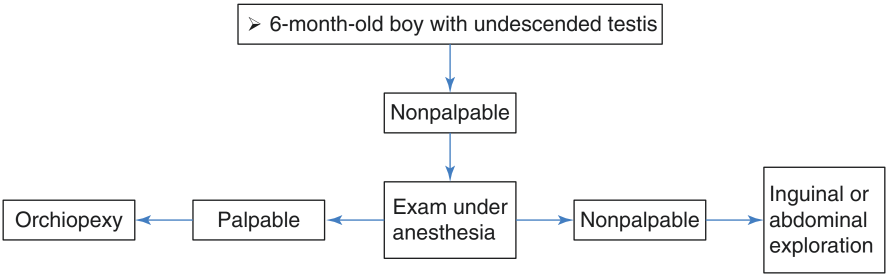
➢ 6-month-old boy with undescended testis
Nonpalpable
Exam under anesthesia
Palpable
Orchiopexy
Nonpalpable
Inguinal or abdominal exploration

**Fig. 1** Management of undescended testes (Source: Adapted from Partin AW, Dmochowski RR, Kavoussi LR, Peters CA, Wein AJ. Campbell Walsh Wein Urology, 12th Edition. Elsevier, Amsterdam, NL, 2020, Chapter 46, Fig 46.6, page 958)

f. Inguinal orchiopexy
   i. Standard approach for a palpable UDT.
   ii. Low transverse incision in inguinal region skin crease. Expose the external ring and incise the external oblique fascia to expose the canal. Isolate the spermatic cord.
   iii. The gubernaculum is divided with cautery.
   iv. If a patent processus vaginalis is present, it must be carefully mobilized off the cord structures and a high ligation is performed.
   v. Do not disturb the veil of tissue between the vas and testicular vessels (Fowler Stephens membrane) to preserve all collaterals.
   vi. All lateral spermatic attachments are divided. If necessary, in higher cases, formal division of all retroperitoneal attachment is accomplished by following the cord retroperitoneally. Careful dissection in the plane between the cord and peritoneal sac is the key. Prentiss maneuver: medial transposition of the testis beneath the epigastric vessel, may be employed.
   vii. Placement of testis in subdartos pouch
      1. Creation of a subdartos pouch through a separate scrotal incision.
      2. Three-point fixation: sutures placed between the tunica vaginalis and dartos fascia to secure testicle orientation. No sutures are placed through the tunica albuginea of the testicle due to risk of vascular injury to the testis.

g. Surgical approach for non-palpable UDT
   i. Diagnostic laparoscopy is a primary option.
   ii. Careful examination under anesthesia to assess for a palpable testicle. For a scrotal nubbin, in the setting of contralateral compensatory growth, scrotal exploration may suffice.
   iii. On laparoscopy, blind-ending vessels indicate the absence of the testis, and no further treatment is needed.
   iv. Spermatic vessels must be located. Occasionally, the vas is seen, and the UDT can be completely separated in a much higher position.
   v. If UDT is intra-abdominal and it can be mobilized into the scrotum, a standard laparoscopic orchiopexy is done.
   vi. If bilateral intra-abdominal UDTs are present, or the UDT is very high in position, consider ligation of the spermatic vessels in a staged Fowler-Stephens procedure. Ligation of spermatic vessels must be done primarily, and not after mobilization of the cord.

h. Laparoscopic Orchiopexy
   i. Bladder and stomach must be empty.
   ii. Mobilize structures distal to the internal ring (epididymis/vas and gubernacular remnant).
   iii. Dissect the peritoneum lateral to the spermatic vessels and distal to the vas.

        iv. Proximally mobilize the vessels while maintaining blood supply to the vas and spermatic vessels.
        v. Do not disturb the veil of tissue between the vas and testicular vessels (Fowler Stephens membrane) to preserve all collaterals.
        vi. Place laparoscopic trocar through the scrotal incision into the abdominal cavity.
        vii. Enter just lateral to the median umbilical ligament.
        viii. Subdartos pouch fixation.
        ix. If there is tension on the cord, sometimes additional length can be gained with further mobilization superior on vessels.

    i. Staged laparoscopic Fowler-Stephens orchiopexy

        i. Wait at least 6 months from initial ligation of vessels to second stage of operation to allow improved collateral vessels to develop.
        ii. 60–70% long-term success.

4. Outcomes

    a. Successful placement of UDT in intrascrotal position via standard inguinal orchiopexy without atrophy is possible in over 98% of cases in experienced hands.
    b. Up to 80–90% success rate is possible after laparoscopic orchiopexy.
    c. Orchidopexy before 8 years of age may mitigate some of the increased risks for testicular cancer, but an increased risk 3–4 times higher than normal persists. The risk of cancer is 8 times higher than normal when the orchidopexy is performed after the onset of puberty. All boys with a history of UDT must receive instruction on self-testicular examination for tumor surveillance with onset of puberty.

5. Prognosis

    a. Fertility

        i. Although semen analysis or testicular biopsies may reveal abnormalities in cases of unilateral UDT, paternity is similar to men born with two descended testicles.
        ii. Paternity falls to 60–65% of normal in cases of bilateral UDT.

    b. Malignancy

        i. Increased risk of testicular germ cell tumor (3–4 times higher). Orchiopexy allows for easier testicular examination for detection of a testicular tumor later in life but does not prevent the risk of malignancy.
        ii. Positively correlated with age.
        iii. Some recommend proceeding with a testicular biopsy in pubertal and postpubertal boys.

## Additional Notes

Cryptorchidism, or undescended testis, is a complex process that is not fully understood. It is typically completed in the third trimester of gestation or after birth. There are likely multiple factors that contribute to failure of testicular descent including genetic and environmental factors.

It is especially important to differentiate cryptorchidism from retractile testes. A thorough physical examination of the child is necessary. Tactics to relax the child may be needed including distraction of the patient, use of warm hands, a warm examination room, or soap or liquid. If a child is difficult to examine, it is important to have the child positioned in numerous positions to best identify the location of the testis. Sometimes, a patient is needed to return for multiple visits to have a thorough examination.

We have seen that many referrals are placed with patients sent with a scrotal ultrasound. It is important to note that cryptorchidism is diagnosed based on a clinical diagnosis. Imaging is not required and should not be utilized. It is important that if there is any concern for cryptorchid testis, a patient should be referred to a specialist for further evaluation.

## Study Questions

1) Question 1: The mother of a 2-month-old infant brings her son to the clinic for a well child check. She is concerned because she feels only one testis in the scrotal sac. Which of the following statements about the next step in management is the most accurate?

   a. Obtain a scrotal ultrasound
   b. Orchiopexy
   c. Return for repeat examination in 4 months
   d. Diagnostic laparoscopy

   Answer to Question 1:

   (c) Return for repeat examination in 4 months. Undescended testis is a clinical diagnosis and does not require any imaging to help guide management. Typically, testicles descend by 6 months of age. If the testicle has not descended into the scrotum, by 6 months of age, surgical management is warranted with orchiopexy. However, since this patient is still <6 months, the best next step is to re-examine the child when he is 6 months.

2) Question 2: A mother of a 6-month-old infant brings her child to the urology clinic for evaluation of bilateral undescended testis. She is concerned as he has always had an empty scrotum. What is the most accurate next step?

a. Obtain serum anti-Mullerian hormone
b. Bilateral orchiopexy
c. Repeat examination in 6 months
d. Diagnostic laparoscopy

Answer to Question 2:

(a) Obtain serum anti-Mullerian hormone. Patients with bilateral nonpalpable testis need to be evaluated for DSD (disorders of sexual differentiation). Referral to a specialist is warranted if there is a concern for bilateral undescended testis. Obtaining a serum anti-Mullerian hormone (AMH) is necessary to guide management. If measurable, surgical exploration is necessary. If AMH is not detected, surgery is unnecessary as the patient is anorchid.

3) Question 3: A 40-year-old patient presents to the clinic. He reports he had an undescended testis that was treated with an orchidopexy when he was 6 months old. He asks the physician if he is at an increased risk for testicular cancer. What is the best response?

a. You are at an increased risk for testicular cancer.
b. You are not at an increased risk for testicular cancer.
c. We should proceed with a testis biopsy.
d. If your orchidopexy was performed after puberty, you would be at lower risk for testicular cancer.

Answer to Question 3:

(a) You are at an increased risk for testicular cancer. Orchidopexy prior to 8 years of age may mitigate some of the increased risks for testicular cancer, but an increased risk 3–4 times higher than normal persists. The risk of cancer is 8 times higher than normal when the orchidopexy is performed after the onset of puberty. All boys with a history of UDT must receive instruction on self-testicular examination for tumor surveillance with onset of puberty. Some recommend proceeding with a testicular biopsy in pubertal and postpubertal boys who have not yet undergone orchidopexy.

## Further Reading

Partin AW, Dmochowski RR, Kavoussi LR, Peters CA, Wein AJ. Campbell Walsh Wein urology. 12th ed. Amsterdam, NL: Elsevier; 2020.

Smith JA, Howards SS, Preminger GM, Dmochowski RR. Hinman's Atlas of urologic surgery revised reprint. 4th ed. Amsterdam, NL: Elsevier; 2019.

Villanueva CA, Dudley AG, Authors, Jacobs MA, Editors. Undescended testis. Linthicum, MD: AUA University, American Urological Association; 2020. https://university.auanet.org/core/pediatric/undescended-testis/index.cfm. Accessed 9 Nov 2021.

# Choledochal Cyst

**Yi-Horng Lee**

**Key Points**

1) Choledochal cyst is either diagnosed by prenatal ultrasound or when it presents with the clinical findings of abdominal pain, palpable mass, and jaundice.
2) The most common type (85%) is type I, which is a fusiform dilatation of the common bile duct.
3) Choledochal cyst requires resection and reconstruction of bile drainage to the small bowel, both to resolve symptoms of biliary obstruction and to prevent future neoplasia which could occur in the wall of the cyst.

Choledochal Cyst is a malformation of the biliary tree. Majority of them are extrahepatic, but some may involve intrahepatic biliary tree. Most cases are clinically silent in childhood, but the majority of them present with abdominal mass, jaundice, or right upper quadrant pain by 10 years of age. If untreated, it can result in cholangitis or malignancy.

1. Pathophysiology
    a. Demographics:
        i. The incidence of choledochal cysts in the Western population is estimated to be 1:100,000 to 1:150,000.
        ii. Increased incidence in Asians: close to ten times more common.
        iii. Female to male ratio is four to one.

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b. Etiology: May be associated with abnormal pancreaticobiliary junction that results in increased pressure and retrograde flow of pancreatic secretions into the common bile duct. This "Common Channel Theory" hypothesizes that mixing of pancreatic and biliary secretions activates the pancreatic enzymes, thus causing inflammation and deterioration of the biliary duct wall [1, 2].

c. Types of Choledochal cyst (Classification by Alonzo-Lej, Todani, and others [3, 4]) (Fig. 1).

   i. Type I: Cystic (fusiform) dilation of the common bile duct, without involvement of the intrahepatic biliary tree (85%).
   ii. Type II: Diverticular malformation of the common bile duct (saccular).
   iii. Type III: Choledochocele within the wall of the duodenum.
   iv. Type IV: Multiple dilations of the biliary tree (10–15%).
      1. Type IVa: Multiple dilations of both intrahepatic and extrahepatic biliary tree.
      2. Type IVb: Multiple dilatations of the extrahepatic bile ducts.

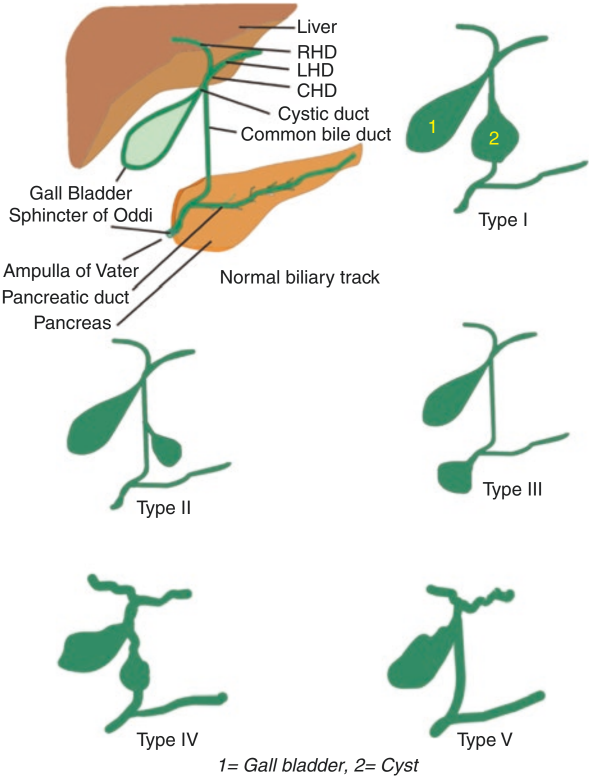
Liver
RHD
LHD
CHD
Cystic duct
Common bile duct
Gall Bladder
Sphincter of Oddi
Ampulla of Vater
Pancreatic duct
Pancreas
Normal biliary track
1
2
Type I
Type II
Type III
Type IV
Type V
1= Gall bladder, 2= Cyst

**Fig. 1** Types of choledochal cyst. Type I: fusiform dilation of the common bile duct. Type II: saccular diverticulum of the common bile duct. Type III: choledochocele within the wall of duodenum. Type IV: intrahepatic and extrahepatic biliary dilation, Type V: intrahepatic biliary dilation. CHD: common hepatic duct, LHD: left hepatic duct, RHD: right hepatic duct. 1: Gallbladder, 2: Choledochal cyst (Source: I, Drriad, Used under Creative Commons Attribution-Share Alike 3.0 Unported License, https://en.wikipedia.org/wiki/Choledochal_cysts#/media/File:Choledochal_cysts.svg, Accessed NOV 11, 2021)

        v. Type V: Multiple biliary cysts of the intrahepatic biliary tree (Caroli disease).
        vi. Type VI: Isolated cyst of the cystic duct.

    d. Caroli disease is a hepatic condition with multiple cystic dilatations of the intrahepatic biliary tree. Caroli syndrome is when these dilatations are associated with hepatic fibrosis and portal hypertension. Caroli disease is associated with Autosomal Recessive Polycystic Kidney Disease and mutations of the PKHD1 gene.
    e. Risk of malignancy is approximately 11% of choledochal cyst [5]. The most common type of malignancy is cholangiocarcinoma [6], but pancreatic and gallbladder cancers may also occur. The incidence of cancer increases with age. Cancer is most commonly associated with types I and IV cysts. Due to risks of cancer, resection is preferable to drainage procedures. When compared to complete cyst excision, drainage of the cyst has four times the risk of malignancy.

2. Clinical presentation

    a. Prenatal diagnosis with sonography has become a more common mode of presentation.
    b. Newborns: Present with obstructive jaundice with or without a palpable mass.
    c. Infants: Present with abdominal pain, abdominal mass, and jaundice.
    d. Adolescents and adults: Vague chronic abdominal pain, mass, and occasional pancreatitis.
    e. Other symptoms may include fever, nausea/vomiting, pruritus, and weight loss.

3. Diagnosis

    a. History:

        i. Jaundice
        ii. Abdominal pain
        iii. Abdominal mass
        iv. History of poor feeding and growth
        v. Acholic stool

    b. Physical: Check for jaundice and abdominal mass
    c. Laboratory:

        i. Complete blood count, electrolytes, liver function tests, pancreatic enzymes.
        ii. Consider other liver diseases such as viral hepatitis and Alpha-1 antitrypsin deficiency.

d. Imaging.
    i. Prenatal sonography is a common way in which choledochal cysts are discovered.
    ii. Postnatal sonography will further characterize the type and location of the cyst(s).
    iii. MRCP reveals the anatomy of the biliary tree. This can help establish the diagnosis and provide insight for preoperative planning.
    iv. HIDA scan will reveal if biliary obstruction is present. It can also differentiate from other causes of neonatal jaundice.
    v. ERCP is useful when biliary obstruction is present and pinpoint the site of obstruction. The obstruction can be relieved by sphincterotomy.
    vi. Intraoperative cholangiography can aid in identification and dissection of branches of the biliary tree during excision of cyst.

4. Treatment
    a. Choledochal cyst is treated with excision.
        i. Cyst must be excised to address or prevent biliary obstruction.
        ii. If not resected, cyst carries a risk of future malignancy, most commonly cholangiocarcinoma.
        iii. Total cyst excision is possible in most cases. However, the inflammation may cause severe scarring, which makes dissection from surrounding structures difficult. In these cases, the cyst is opened, and the lining of the cyst is excised via an intramural plane or cauterized.
        iv. After excision, drainage of bile from the liver must be re-established.
        v. In severe Caroli disease, liver transplantation needs to be considered.
    b. Methods of reconstruction of biliary tree after cyst excision.
        i. Roux-en-Y hepatico- or choledocho-jejunostomy depending on the location of the cyst.
        ii. Choledochoduodenostomy is possible, but less preferable due to increased incidence of cholangitis.

5. Outcome
    a. Most patients do well with early cyst excision
    b. Potential surgical complications
        i. Anastomotic leak
        ii. Cholangitis
        iii. Biliary obstruction
    c. Patients with intrahepatic disease will have progressive dilation of intrahepatic cysts and ductal ectasia.
    d. The epithelial lining of the unresected portion of the cyst can undergo malignant degeneration and develop cholangiocarcinoma.
    e. There are also reports of cholangiocarcinoma after a cyst has been excised.

## Additional Notes

Choledochal cyst is a congenital condition that is likely to be encountered in any busy pediatric surgery practice. The increasing use of prenatal ultrasound screening has allowed early detection of many choledochal cysts before the onset of symptoms. This diagnosis needs to be considered in any abdominal mass seen during a prenatal screening ultrasound.

Once a choledochal cyst is detected, resection of the cyst is indicated to resolve or prevent symptoms such as jaundice, cholangitis, or abdominal pain. More importantly, it will markedly decrease the risk of future malignancy. Resection rather than drainage is almost mandatory. Roux-en-Y reconstruction to restore bile flow after resection is preferred due to lower incidence of cholangitis.

There are advances in surgical techniques such as laparoscopic or robotic approaches to this disease. 4K video imaging has enhanced intraoperative visualization and thereby reduced operative time significantly [7]. IV indocyanine green cholangiography has replaced traditional intraoperative cholangiogram.

Postoperative abdominal sonography can be used to evaluate for postoperative complications such as biliary obstruction, pancreatitis, pancreatic fistula, bowel obstruction, and intussusception [8].

Pathologic examination of the resected specimen will diagnose existing malignancy if present. Postoperative surveillance for malignancy is needed all patients throughout their lives.

## Study Questions

1) Question 1: Which is the most common type of choledochal cyst?

   a. Type I: Cystic (fusiform) dilation of the common bile duct, without involvement of the intrahepatic biliary tree.
   b. Type II: Diverticular malformation of the common bile duct (saccular).
   c. Type III: Multiple dilations of biliary tree.
   d. Type IV: Multiple dilations of biliary tree.

Answer to Question 1:

(a) Type I: Cystic (fusiform) dilation of the common bile duct, without involvement of the intrahepatic biliary tree. A Type I choledochal cyst is the most common type of choledochal cyst, representing 90% of all children presenting with this disease. This type is amenable to curative resection and reconstruction of biliary drainage to the intestinal tract with a Roux-en-Y loop of jejunum anastomosed to the stump of the common hepatic duct or the porta hepatis because there is not intrahepatic disease.

2) Question 2: Which statement is not true about malignancy arising from a choledochal cyst?
   a. Median age to develop malignancy is 50 years of age.
   b. The most common malignancy that occurs in cholangiocarcinoma.
   c. Resection of the choledochal cyst has a lower incidence of malignancy than simple drainage of the cyst.
   d. Resection of the choledochal cyst should be delayed until malignancy has been identified.

Answer to Question 2:

(d) Resection of the choledochal cyst should be delayed until malignancy has been identified. This statement is false. The choledochal cyst should be resected at the time it is diagnosed. Often when malignancy is identified, it has already undergone spread and is unresectable. Cyst resection performed early serves to prevent the development of future malignancy. Cyst presentation is usually in the neonatal period or infancy, and median age of developing malignancy is 50 years of age. Cholangiocarcinoma is the most common cancer arising in a choledochal cyst. Simple drainage of a cyst has a fourfold increase in incidence of malignancy when compared to cyst excision and reconstruction of bile drainage.

3) Question 3: Which hepatobiliary condition is characterized by cystic dilatation of the intrahepatic bile ducts?
   a. Choledochal cyst
   b. Caroli disease
   c. Viral hepatitis
   d. Alpha-1 anti-trypsin deficiency

Answer to Question 3:

(b) Caroli disease. The differential diagnosis of cystic malformation of the biliary tree includes Caroli's disease. vii. Caroli disease is a hepatic condition with multiple cystic dilatations of the intrahepatic biliary tree. Caroli syndrome is when these dilatations are associated with hepatic fibrosis and hypertension. Caroli disease is associated with Autosomal Recessive Polycystic Kidney Disease and mutations of the PKHD1 gene.

## References

1. Babbitt DP. Congenital choledochal cysts: new etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. Ann Radiol (Paris). 1969;12(3):231–40.
2. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts. Part 1 of 3: classification and pathogenesis. Can J Surg. 2009;52(5):434–40.
3. Todani T, Watanabe Y, Toki A, Moritomi Y. Classification of congenital biliary cystic disease: Special reference to type Ic and IVA cysts with primary ductal stricture. J Hepato-Biliary-Pancreat Surg. 2003;10:340–4.

4. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg. 1977;134(2):263–9.
5. Ten Hove A, de Meijer VE, Hulscher JBF, de Kleine RHJ. Meta-analysis of risk of developing malignancy in congenital choledochal malformation. Br J Surg. 2018;105(5):482–90.
6. Bismuth H, Krissat J. Choledochal cystic malignancies. Ann Oncol. 1999;10(Suppl. 4):S94–8.
7. Koga H, Ochi T, Seo S, Miyake Y, Lane GJ, Yamataka A. Comparison of 2K and 4K imaging systems for laparoscopic repair of choledochal cyst in children. J Pediatr Surg. 2022;57(2):235–8. https://www.sciencedirect.com/science/article/abs/pii/S0022346821007570.
8. Hosokawa T, Hosokawa M, Shibuki S, Tanami Y, Sato Y, Ishimaru T, Kawashimi H, Oguma E. Role of ultrasound in follow-up after choledochal cyst surgery. J Med Ultrason. 2021;48:21–9.

# Gastroesophageal Reflux

**Benjamin Kuhn and Kritika Sukumar**

**Key Points**

1) Uncomplicated Gastroesophageal Reflux Disease (GERD) may be treated conservatively with lifestyle modification.
2) Warning signs may indicate underlying non-GERD pathological processes.
3) Anti-reflux surgery is reserved for children with severe GERD refractory to medical management.

1. **Epidemiology:**
   a. Prevalence: GERD symptoms are present in 40% of young infants and show a steady decline with increasing age. In older children, GERD symptoms are present in 10% of children.
   b. GER: Gastroesophageal Reflux (uncomplicated gastroesophageal reflux) is the effortless retrograde passage of gastric contents into the esophagus.
   c. GERD: Gastroesophageal Reflux Disease (complicated gastroesophageal reflux) is the symptoms or complications that may occur when gastric contents reflux into esophagus.
2. **Pathophysiology:**

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   a. The primary mechanism of GERD is transient lower esophageal sphincter (LES) relaxation.
   b. Secondary mechanisms include increased intraabdominal pressure, delayed gastric emptying, decreased gastric compliance, and decreased esophageal capacitance.
   c. Risk Factors (all ages):
      i. Alcohol
      ii. Hiatal Hernia
      iii. Hypotonia
      iv. Motility Disorders
      v. Obesity
      vi. Premature Birth
      vii. Psychological factors: stress, anxiety, depression
      viii. Smoking

3. **Clinical features:**
   a. Infantile GERD can present as excessive crying, irritability, arching of back during feeds, recurrent vomiting, feeding refusal, poor weight gain, and sleep disturbances.
   b. In older children and adolescents, GERD more commonly presents as heartburn, retrosternal chest pain, or regurgitation sensations.

4. **Diagnosis:**
   a. In most cases of GERD, a thorough history and physical examination are sufficient for diagnosis. Refer to Fig. 1 Algorithm.
   b. Diagnostic imaging should be utilized as needed for atypical presentation and to rule out complications, predisposing conditions, or alternative diagnoses.
      i. *Upper GI Contrast Study:* A radiographic study with barium ingestion to evaluate for structural abnormalities of the upper gastrointestinal tract, such as malrotation, hiatal hernia, or strictures.
      ii. *pH Impedance Probe:* A thin flexible catheter measuring acidic and non-acidic reflux events over a 24-h period.
      iii. *Gastric Emptying Scan:* A functional study evaluating the passage of gastric contents into the small bowel to assess for gastroparesis, which can contribute to GERD.
      iv. *Esophagogastroduodenoscopy (EGD):* To evaluate for endoscopic and histopathologic changes of esophageal mucosal injury and inflammation. Useful to evaluate for Eosinophilic Esophagitis which can mimic GERD.

5. **Treatment:**
   a. The primary goal of treating GERD is aimed at reducing symptoms, improving quality of life, and preventing complications of GERD. In uncomplicated infantile GERD, conservative measures such as smaller and more frequent

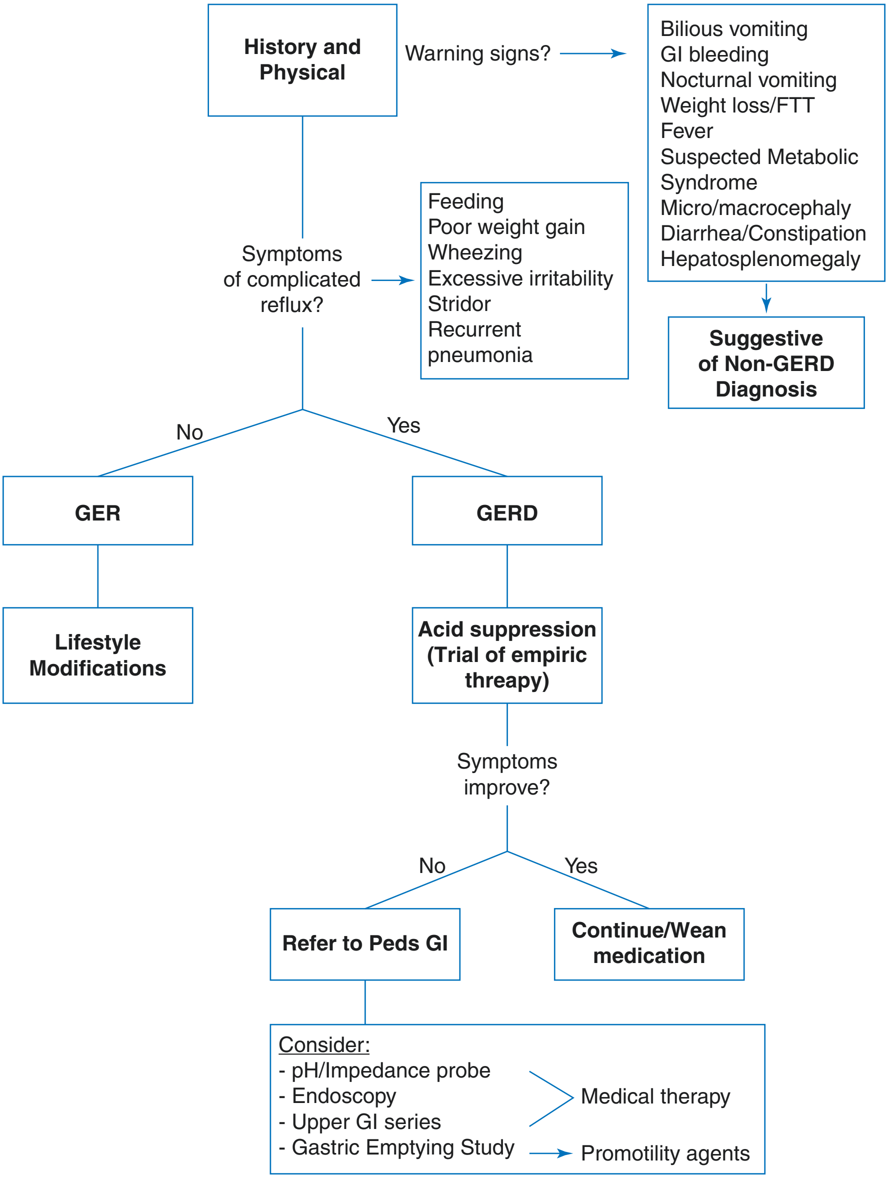
History and Physical
Warning signs?
Bilious vomiting
GI bleeding
Nocturnal vomiting
Weight loss/FTT
Fever
Suspected Metabolic Syndrome
Micro/macrocephaly
Diarrhea/Constipation
Hepatosplenomegaly
Suggestive of Non-GERD Diagnosis
Symptoms of complicated reflux?
Feeding
Poor weight gain
Wheezing
Excessive irritability
Stridor
Recurrent pneumonia
No
Yes
GER
GERD
Lifestyle Modifications
Acid suppression (Trial of empiric threapy)
Symptoms improve?
No
Yes
Refer to Peds GI
Continue/Wean medication
Consider:
- pH/Impedance probe
- Endoscopy
- Upper GI series
- Gastric Emptying Study
Medical therapy
Promotility agents

**Fig. 1** Algorithm 1. Diagnostic approach to pediatric gastroesophageal reflux disease. Source: Kritika Sukumar, MD and Benjamin Kuhn, DO

feeds, thickened formula, and postprandial upright positioning may be sufficient to diminish regurgitation.

b. If these conservative measures fail, pharmacological therapy with gastric acid suppressants may be appropriate. In older children and adolescents, dietary modifications, weight management, positional changes, avoidance of alcohol, and cessation of smoking should be advocated.

c. Medications:

   i. H2 Antagonists: Type 2 Histamine is one of several acid-producing stimuli in the stomach. H2 Antagonists block this receptor.
   ii. Proton Pump Inhibitors (PPI): Block acid production at the source, the proton pump, making this superior to H2 Antagonists in that it reduces gastric acid production across all stimuli (not only the H2 receptor).
   iii. Promotility Agents: Stimulate peristalsis, thereby enhancing gastric emptying.

d. Operative Treatment:

   i. Anti-reflux surgery is reserved for children with severe GERD refractory to medical management, patients who develop complications such as failure to thrive and respiratory symptoms, and some patients with neurological impairment.
   ii. Fundoplication is the most common anti-reflux procedure and involves partial or complete wrapping of stomach fundus around the esophagus above the gastroesophageal junction. Fundoplication prevents reflux episodes by decreasing the number of transient lower esophageal sphincter relaxations and increasing the lower esophageal sphincter pressure.
   iii. This procedure can be performed open or laparoscopically. Major complications following fundoplication include breakdown of the wrap, intrathoracic herniation of the wrap, bowel obstruction, gas bloat syndrome, dysphagia, early satiety, dumping syndrome, and postoperative retching and gagging.

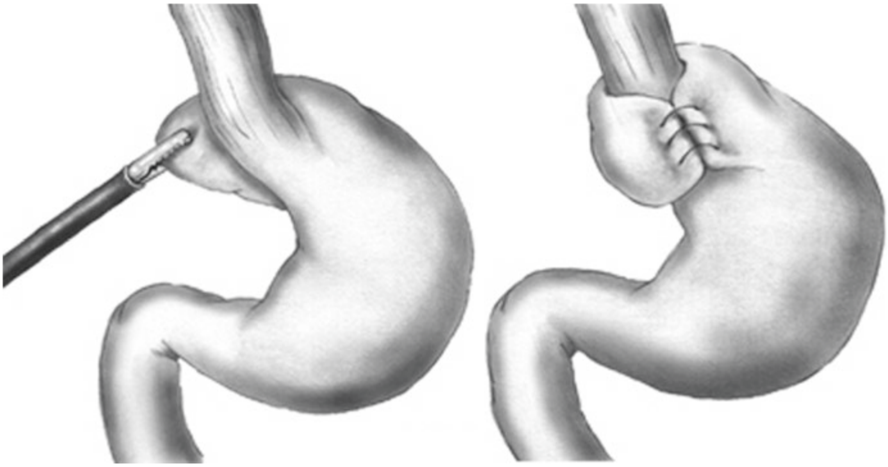

**Fig. 2** Nissen Fundoplication. In the Nissen fundoplication procedure, the fundus of the stomach is used to create a 360° wrap around the lower esophagus (Source: Dr. James P. Gray, released to public domain, via https://upload.wikimedia.org/wikipedia/commons/d/dc/Nissen_fundoplication.png. Downloaded on Dec 24, 2013)

iv. The most commonly performed operation for GERD is the Nissen fundoplication 360° wrap of the gastric fundus around the esophagus (see Fig. 2).

## Additional Notes

There is a growing body of evidence on the negative effects of suppressing gastric acid. The digestion of protein benefits from stomach acid conversion of pepsinogen (zymogen form) into activated pepsins (enzymatic form). The pepsin proteinases break down food proteins into oligopeptides and amino acids, which can then be digested and absorbed into the small bowel. When this process is impaired, there is potential for reduced absorption as well as potential for immune response. In epidemiologic studies, the use of acid suppression in infants has been associated with the later development of IgE-mediated food allergies as well as the development of eosinophilic esophagitis. Long-term use of acid suppression can also impair the absorption of some vitamins and minerals, such as magnesium. Strongly suppressing gastric acid also removes a layer of defense against enteric pathogens. Acid suppressant use is associated with a higher likelihood of developing clostridium difficile infection when compared to those not taking acid suppressants.

As such, current practice patterns for the treatment of reflux symptoms within pediatrics trends toward conservative measures with optional medication trials. If medications are improving symptoms, then the physician should plan a treatment course strategy including an attempted taper off medication as tolerated.

## Study Questions

1. Question 1: A 6-month-old female presents for her well child visit with concerns of regurgitation with all feeds. Patient has normal development and her weight has remained at the 15th percentile. The mother reports occasional back arching with feeds, intermittent irritability, and spits up which resemble curdled milk.

   The best initial management of this child would be:

   A. Start an H2 blocker therapy
   B. Start a proton pump inhibitor therapy
   C. Administration of small, thickened oral feedings
   D. Consider switching to an elemental formula
   E. Referral for a Nissen Fundoplication

Answer: (c.) Administration of small, thickened oral feedings. This is an example of uncomplicated gastroesophageal reflux. This patient does not endorse any alarm symptoms including signs of weight loss or lack of weight gain. Initial treatment would be to initiate lifestyle changes such as administering smaller volume feeds, thickening feeds with rice cereal, and keeping infants positioned upright after feeds.

2. Question 2: A 6-month-old infant presents with irritability and frequent emesis that is described as bilious.

   The best initial management for this child would be:

   A. Start H2 Antagonist Medication
   B. Start Proton Pump Inhibitor Medication
   C. Administer smaller, more frequent feedings with rice cereal thickening
   D. Evaluate for underlying anatomical abnormalities
   E. Referral for fundoplication anti-reflux surgery

Answer: (d.) Evaluate for underlying anatomical abnormalities. Vomiting (forceful expulsion of gastric contents from the mouth), particularly when bilious, may indicate underlying anatomical pathology, such as malrotation, which is a surgical emergency.

3. Question 3: A 3-year old severely developmentally delayed male presents with recurrent episodes of aspiration pneumonia that occur following reflux events.

   The best initial management for this child would be:

   A. Start H2 Antagonist Medication
   B. Start Proton Pump Inhibitor Medication
   C. Administer smaller, more frequent meals
   D. Refer for fundoplication anti-reflux surgery

Answer: (d.) Refer for fundoplication anti-reflux surgery. This child has GERD and suffers from its complications. Especially in neurologically devastated conditions with abnormal oromotor coordination, fundoplication is an appropriate treatment approach.

## Further Reading

Kuhn BR, Young AJ, Justice AE, Chittoor G, Walton NA. Infant acid suppression use is associated with the development of eosinophilic esophagitis. Dis Esophagus. 2020;33(10):doaa073. https://doi.org/10.1093/dote/doaa073.

Rosen R, Vandenplas Y, Singendonk M, Cabana M, DiLorenzo C, Gottrand F, Gupta S, Langendam M, Staiano A, Thapar N, Tipnis N, Tabbers M. Pediatric gastroesophageal reflux clinical practice guidelines: joint recommendations of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition. J Pediatr Gastroenterol Nutr. 2018;66(3):516–54. https://doi.org/10.1097/MPG.0000000000001889.

Singendonk M, Goudswaard E, Langendam M, van Wijk M, van Etten-Jamaludin F, Benninga M, Tabbers M. Prevalence of gastroesophageal reflux disease symptoms in infants and children: a systematic review. J Pediatr Gastroenterol Nutr. 2019;68(6):811–7.

Trikha A, Baillargeon JG, Kuo YF, et al. Development of food allergies in patients with gastroesophageal reflux disease treated with gastric acid suppressive medications. Pediatr Allergy Immunol. 2013;24(6):582–8.

# Benign Disorders of the Anus

**Neesha S. Patel, Steven R. Donahue, Katharine A. Armstrong, Kelsey R. Tieken, and Angela M. Hanna**

**Key Points**

1. Understand normal anatomy of the anus and rectum.
2. Recognize some of the most common benign pediatric disorders of the anus as anal fissures, hemorrhoids, rectal prolapse, abscess, juvenile polyps, dyssynergic defecation, congenital perianal masses, anorectal malformations, etc.
3. Be able to differentiate disorders that often present with common complaints.
4. Be able to describe when surgery is indicated and what medical therapies are available.
5. Understand basic surgical principles and management of the anus.

## Anal Fissure

1. Background
    a. Anal fissures are often seen in children.

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   b. Fissures are a common problem in pediatric patients; however, the incidence is difficult to establish because anal discomfort is often misattributed to other etiologies.

2. Pathophysiology

   a. The most common underlying etiology is constipation; passage of a large, firm stool causes a longitudinal tear in the anoderm resulting in a primary fissure. These generally occur in the posterior midline; however, a small percentage may occur in the anterior midline. Anterior midline fissures are more common in females.
   b. Hypertonia of the internal sphincter or spasm then occurs, which impairs blood flow to the injured area and slows healing. Secondary fissures may also occur and should prompt further workup for signs of infection or trauma. Fissures may be acute, which are generally more painful but heal within a few days to weeks, or chronic, which are present for more than 6 weeks but often present with less pain. Chronic fissures may be difficult to heal.

3. Clinical Presentation

   a. Patients with a fissure typically present with pain or crying with defecation, rectal bleeding, or stool with red streaks around it.

4. Diagnosis

   a. The diagnosis is confirmed with an exam and if the patient cannot tolerate the exam while awake, it may require anesthesia. For atypical fissures or if multiple fissures are found, screen for other etiologies such as infection, trauma, or sexual abuse. Multiple recurrent fissures may also be a sign of inflammatory bowel disease.

5. Treatment

   a. Initial management of fissures focuses on relieving the inciting factors. Focus on increasing water and fiber intake and consider adding a bowel regimen such as docusate or polyethylene glycol and investigate the underlying cause of constipation.
   b. Topical anesthetic agents can be used for symptom relief. Most fissures will resolve with conservative management alone.
   c. Second-line treatment for persistent fissures consists of topical calcium channel blockers (diltiazem or nifedipine cream/gel) or nitroglycerin. Side effects to these medications are minimal, but vasodilatory agents can cause headaches. It is important to recognize that while these agents are effective, they are never used alone.
   d. Continued treatment of constipation is key to successful resolution of the fissure.

   e. Third-line treatment is botulinum toxin injections which cause sphincter relaxation. Sphincterotomy is not recommended in young children due to the thin nature of the developing muscle complexes in which disruption can cause incontinence.

## Solitary Rectal Ulcer Syndrome

1. Background
   a. This is a benign chronic disorder in children related to constipation and excessive straining. It is an uncommon disorder and is better reported in adults rather than children. If it presents in childhood, then the average onset is around 3 years of age.
2. Pathophysiology
   a. It results from rectal hypersensitivity and leads to a persistent desire to defecate and a sensation of incomplete emptying. Ultimately this leads to continuous internal rectal prolapse.
3. Clinical Presentation
   a. First symptoms are rectal bleeding, passage of mucus while defecating, tenesmus, perineal and lower abdominal pain, feelings of incomplete defecation, and rectal prolapse.
4. Diagnosis
   a. Diagnosis is made with a thorough history and physical. Underreporting is common due to the nonspecific symptoms.
   b. Defecography can show intussusception, internal/external mucosal prolapse, non-relaxing puborectalis, and incomplete/delayed emptying.
   c. Endoscopies will show shallow ulcerating lesions surrounded by hyperemic mucosa, most commonly on the anterior side of the rectum.
   d. Biopsies are necessary for definitive diagnosis and show fibromuscular obliteration of lamina propria and disorientation of muscle fibers.
   e. Endorectal ultrasound shows a thick internal anal sphincter.
   f. Other chronic diseases must be ruled out, such as IBD, amebiasis, chronic ischemic colitis, endometriosis, colitis cystica profunda, and malignancy.
5. Treatment
   a. Treatment is geared toward constipation—more water and fiber intake, regular enemas, laxatives, regulation of bowel habits with decreased straining.
   b. Biofeedback and behavioral modifications are effective if children are compliant.
   c. Surgical options only if prolapse is present and includes rectopexy, mucosal excision, Delorme’s procedure.

## Hemorrhoids

1. Background
    a. Symptomatic hemorrhoids are one of the most common anorectal complaints in children, but incidence is low overall. In young children, the most common cause is liver failure and in adolescents they are caused by chronic constipation.
2. Pathophysiology
    a. Hemorrhoids are the vascular subcushions that exist in the anal canal at the left lateral, right anterior, and right posterior positions. When these submucosal cushions become enlarged with venous congestion, the connective tissue around the anus loosens and they may protrude from the anus.
    b. Internal hemorrhoids are graded based on the degree of prolapse:
        i. Grade I hemorrhoids bulge into the anal canal with no prolapse.
        ii. Grade II prolapse but will spontaneously reduce.
        iii. Grade III requires manual reduction of prolapsed tissue.
        iv. Grade IV cannot be reduced.
3. Clinical presentation
    a. These present as soft tissue growth, or bulge, around the anus. Patients may also present with bleeding, itching, prolapse, or pain.
    b. External hemorrhoids arise from below the dentate line and are innervated by somatic nerves, and therefore are frequently painful. A thrombosed external hemorrhoid presents as a firm, tender blue mass. The pain improves after two to three days and hemorrhoid regresses in one to two weeks and may leave a skin tag after resolution. If the pain has begun to improve by the time of presentation, there is no indication for surgical correction, and these can be managed conservatively.
    c. Internal hemorrhoids are innervated by visceral nerves above the dentate line and therefore present with painless bleeding and prolapse.
4. Diagnosis
    a. Diagnosis is confirmed with a physical exam and, if needed, anoscopy. Digital rectal exam is necessary to assess sphincter tone and to rule out other causes of bleeding such as a rectal polyp or a mass.
5. Treatment
    a. Initial treatment is conservative management with treatment of constipation, increased water and fiber intake, aggressive bowel regimens, sitz baths, and prevention of long periods of time spent straining on the toilet. With time and appropriate management, most hemorrhoids will resolve without need for surgical intervention.

   b. If symptoms do not resolve with conservative management, procedural intervention may be required. Consider referral to a colorectal surgeon if excision is required.
   c. Thrombosed external hemorrhoids can be excised in the first 48–72 h, however, after this there is little benefit to excision as pain usually begins to resolve. It is not recommended to rubber band ligate external hemorrhoids because it is extremely painful.
   d. Grades I and II internal hemorrhoids can often be rubber band ligated successfully. For grades III and IV hemorrhoids, or when medical management fails, surgery is the next line of treatment.
   e. Hemorrhoidectomy can be performed open, with a staple device or a LigaSure device (Medtronic, Minneapolis, MN). In younger children, a stapler may be difficult due to size but can be successful in adolescents. Stapled and LigaSure techniques have less complications compared to open techniques, including less pain and decreased risk of stricture or abscess formation.

## Rectal Prolapse (RP)

1. Background
   a. Children are particularly prone to prolapse due to anatomic considerations. The rectum sits lower in the pelvis and takes a more vertical course. The pelvic floor has weak levator muscle support, and the coccyx is relatively flat. In addition, the sigmoid colon is fairly mobile, the rectal mucosa is redundant, and up to 75% of infants less than 1-year old have absent Houston's valves.
2. Pathophysiology
   a. Rectal prolapse is the extrusion of some or all of the rectal mucosa through the external anal sphincter. There are two types. Type I is false procidentia or mucosal prolapse, which is less than 2 cm of prolapse with radial folds at the junction of the mucosa and anus. Type II is true procidentia or rectal prolapse with concentric folds of the mucosa visible protruding through the anus. Type II is further subdivided into different types based on length of protrusion. Rectal prolapse is functionally an intussusception of the rectum.
   b. The most common cause in the USA is chronic constipation. Other, less common causes include increased colonic motility (e.g., infectious diarrhea or laxative abuse), increased abdominal pressure (e.g., constipation, coughing, vomiting, or straining to urinate), and congenital conditions (cystic fibrosis, myelomeningocele, Hirschsprung's disease, spina bifida, or polyps).
3. Clinical presentation
   a. Clinically, rectal prolapse presents as a dark red mass with or without mucus or blood that will protrude from the rectum while straining. While this may cause some mild discomfort to the patient, it is usually painless.

4. Diagnosis

    a. The diagnosis can be made by history alone if parents are able to give a detailed explanation or photographic evidence of prolapse. RP may often not be seen in clinical settings, however, if it is present on physical examination that is enough for definitive diagnosis. The clinician may encourage parents to take videos or photos when this occurs at home as this can prevent unnecessary procedures or examinations.
    b. When working up a child for rectal prolapse, it is important to evaluate for potential underlying causes. A colonoscopy may be performed if there's suspicion of other lesions such as polyps. Defecography can also be useful but is generally not as successful in children compared to adults. MRI is another option but is often difficult in pediatric patients due to the length of the exam. A CF test is warranted only if the patient has recurrent rectal prolapse and has failed medical treatments.

5. Treatment

    a. Several treatment options exist for rectal prolapse. Prolapse in young children is usually a self-limiting disorder and patients tend to improve with behavioral modifications alone. If RP is encountered in the clinical setting, manual reduction of the prolapse should be performed quickly. The child should be placed in a knee-chest position, and sedation can be used if necessary. You can use table sugar to allow an osmotic shift of fluid making it easier to reduce. It is important to send a bowel regimen home with parents after reduction. Always consider underlying causes to treat.
    b. Local therapy is a potential option for RP and includes sclerotherapy and cerclage.
        i. Sclerotherapy requires general sedation and consists of an injection of sclerosant just above the dentate line. This causes fibrosis of the rectal wall to surrounding tissue causing adherence. Ethyl alcohol is typically used as a sclerosant as it results in the least amount of complications and recurrences.
        ii. Cerclage (Thiersch's) is another option which involves a circumferential stitch around anal opening and tying over Hagar dilators. A combination of both sclerotherapy and cerclage can also be used. Both can be complicated by anal pain, stricture, perianal abscesses, and fistulas. Both have high recurrence rates which necessitate repeat procedures. It is important to note that these should not be used in patients with full-thickness RP.
    c. Surgical treatment of rectal prolapse is another potential option. Surgery is typically indicated for older children who have failed medical therapy or are likely to fail. It is not uncommon to have an adult colorectal surgeon involved with the procedure. The following are the most common procedures performed in North America according to the American Pediatric Surgical Association (APSA). Due to the rarity of the condition in children, not much is known about which technique has a better outcome.

    i. Transabdominal rectopexy is typically performed as a laparoscopic suture rectopexy (LSRP) in which the perirectal tissues are sutured to the sacral promontory. A sigmoid rectopexy may also be performed by suturing the sigmoid colon to the left lateral peritoneum. Benefits to this include decreased length of stay (usually an outpatient procedure), less pain, and low morbidity. Similarly, one may perform a transabdominal sigmoidectomy and rectopexy which constitutes the most durable repair. This requires an abdominal procedure and may be performed laparoscopically.
    ii. Another option is a perineal proctectomy or proctosigmoidectomy. Because this involves a perineal approach, the morbidity associated with an abdominal procedure is avoided. Two perineal procedures are described. In an Altemeier procedure, a full-thickness circumferential incision is made in the prolapsed rectum about 1–2 cm proximal to the dentate line. The rectum is delivered, and the mesentery is ligated until it cannot be pulled down any further. The bowel is then transected, and the edge is sewn to the incision. A Delorme procedure involves a mucosectomy of the rectum. The remaining muscular layer is then sutured above the dentate line.
    iii. A posterior sagittal rectopexy may also be performed. In this procedure, a posterior sagittal incision is made. The upper part of the sphincters are then divided, and the rectal wall is defined. This is then sutured to the sacral promontory. This approach eliminates the need for an abdominal incision. However, the outcome is typically not as good if the intussusception of the rectum is higher.

## Perianal Abscess

1. Background/Pathophysiology
    a. Perianal abscesses originate in glands of Morgagni and have a male predominance. There is some evidence that excessive androgens or androgen-sensitive glands are associated. Perianal abscesses most commonly occur in the 3 o'clock and 9 o'clock positions. 20–85% of these can progress to fistula-in-ano. In older children, a perianal abscess may be a warning sign of a diagnosis of inflammatory bowel disease.
2. Clinical presentation
    a. Perianal abscesses present as firm or fluctuant masses located near the anus and often present with pain, irritability, and fever. While the child may appear ill, it is very rare for a child to become septic from these abscesses.

3. Diagnosis
    a. While the condition can be diagnosed on external exam alone, ultrasonography can help define a cavity. However, ultrasonography is not always reliable in predicting the amount of fluid present.
4. Treatment
    a. Several different treatment options exist for perianal abscesses. For children less than 1 year of age, these abscesses will often resolve spontaneously. A conservative approach with antibiotics and warm soaks may be attempted. However, most older children will undergo incision and drainage of the abscess. Antibiotics normally are not indicated unless there were systemic symptoms, or the child has a predisposing condition that would make them immunocompromised.

## Juvenile Polyps

1. Clinical presentation
    a. Juvenile polyps typically present between ages 2–5 years, and the first sign is painless rectal bleeding. These can be mistaken for hemorrhoids if seen at the anal verge.
2. Diagnosis
    a. When noticed, they warrant colonoscopic evaluation of the entire colon. For definitive diagnosis and possible treatments, please follow treatment guidelines for colonic polyps.
    b. Pediatric patients with polyps occurring in early adolescence, polyps outside of the colon, family history of cancer, or with multiple polyps should be evaluated for possible associated syndromes (Juvenile polyposis, Peutz-Jeghers, familial adenomatous polyposis).
3. Treatment
    a. Polypectomy can be performed at the time of colonoscopy/anoscopy, and further treatment algorithms pursued based on final pathology.

## Internal Anal Sphincter Achalasia (Formerly Ultra-Short Hirschsprung's Disease)

1. Background
    a. Internal anal sphincter achalasia is similar in nature to Hirschsprung's disease. It accounts for around 4.5% of children with chronic constipation.

2. Pathophysiology
    - a. The condition is multifactorial in nature but likely secondary to defective innervation of the nitrogen receptors in the internal anal sphincter. The internal anal sphincter normally relaxes in response to distension which causes a release of nitric oxide and ultimately smooth muscle relaxation. Defects along this pathway lead to impaired relaxation and difficulty passing stool.
3. Clinical presentation
4. Diagnosis
    - a. The diagnosis of internal anal sphincter achalasia can be made with anorectal manometry showing the absence of the retrosphincteric reflex after balloon dilation. In contrast to Hirschsprung's disease, rectal suction biopsies will show presence of ganglion cells.
5. Treatment
    - a. Several treatment modalities exist for internal sphincter achalasia. Regular bowel regimens can help but often do not provide adequate symptom relief for these patients. Posterior internal sphincter myectomy may be performed with 5–10 mm wide resection of smooth muscle posteriorly. It is important to achieve the right amount of resection as too little tissue resected leads to continued constipation and too much leads to incontinence.
    - b. Alternatively, intersphincteric Botox (Allergan, Cincinnati, OH) injections may be used. Botox inhibits the release of acetylcholine and denervates the internal sphincter. While this is an effective treatment for achalasia, it only lasts for a transient period.

## Dyssynergic Defecation

1. Background
    - a. Dyssynergic defecation is a functional outlet disorder also known as pelvic floor dyssynergia.
2. Pathophysiology
    - a. Dyssynergic defecation results from ineffective relaxation of the puborectalis and external anal sphincter muscles along with increased intra-abdominal pressures causing chronic constipation. It can also be secondary to inappropriate contraction of the pelvic floor muscles leading to a narrowing of the canal.
3. Clinical presentation
    - a. Children will often present with chronic constipation and associated sequelae of hemorrhoids, fissures, etc.

4. Diagnosis
    a. The diagnosis is made based on anal manometry and balloon expulsion test. On manometry, less than 20% relaxation of the basal resting sphincter tone is seen, and on balloon expulsion test, adequate propulsive force during defecation is noted.
5. Treatment
    a. First-line treatment for dyssynergic defecation is pelvic floor physical therapy along with healthy bowel hygiene.

## Congenital Perianal Masses

Congenital perianal masses include benign acrochordon (skin tag), hamartomatous polyp, and sacrococcygeal teratoma (can present as perianal mass). If considering one of these diagnoses, obtain ultrasound or possibly MRI (not discussed in detail in this chapter, please refer to treatment of specific condition for further details).

## Anorectal Malformations

1. Background/pathophysiology
    a. Congenital rectal atresia and anal stenosis fall on the spectrum of anorectal malformations and are rarely encountered. Patients often have a defective or incomplete anal canal but intact sphincter complexes. Long-term function varies based on the type of defect present.
2. Clinical presentation
    a. These patients typically present with a normal anal opening but failure to pass meconium.
3. Diagnosis
    a. Diagnosis is made by inability to pass a dilator into the anal canal. Because these malformations are often associated with presacral defects and masses, patients require spinal ultrasound/MRI prior to repair.
4. Treatment
    a. In cases with a long atretic segment, it can be difficult to approach from a transanal incision and a posterior sagittal or laparoscopic approach may be used. In general, the goal is to preserve the dentate line and sphincter complex while restoring the anal canal.

## Study Questions

1. A 4-year-old female is brought to the clinic by her mother. The mother states her child has been complaining of anal pain over the past week or two. She has had issues with constipation for some time and yesterday had her first bowel movement in days. When she went to wipe, there were streaks of blood on the toilet paper which prompted the visit today. You are concerned the patient may have an anal fissure, and your working diagnosis is confirmed on exam under anesthesia. What is the next best step in management of this patient?

   a) Perform lateral internal sphincterotomy
   b) Botulinum toxin injection
   c) Excision of the fissure tract
   d) Increase water and fiber intake and consider bowel regimen

Answer: (d.): Increase water and fiber intake and consider bowel regimen. This patient has an anal fissure which is likely secondary to her history of chronic constipation. The first step in management is to treat the underlying cause of the fissure, namely constipation. First-line treatment involves making sure the patient has adequate water and fiber intake. Additionally, you can consider adding a bowel regimen with docusate or polyethylene glycol. Botulinum toxin is second-line treatment. Lateral sphincterotomy is not recommended in children as it can result in permanent incontinence.

2. A 10-year-old boy is brought to the ER by his parents. He has been having anal pain over the past two days and low-grade fevers. On exam, there is a warm, red, tender, fluctuant area at the 3 o'clock position of the anal opening, approximately 3 cm in size. What is the next best step?

   a) Prescribe antibiotics. Follow-up in one week.
   b) Recommend soak in Sitz bath twice daily.
   c) Perform drainage with a scalpel in office under local anesthesia.
   d) Incision and drainage in the operating room.

Answer: (d.): Incision and drainage of the abscess in the operating room. While a conservative approach with antibiotics and warm soaks could be attempted, an abscess in an older child will typically warrant drainage as it is less likely to resolve on its own. Antibiotics should not be necessary after drainage as this child is having only mild fevers without any other systemic symptoms and is immunocompetent. Given the child's age, screening for inflammatory bowel disease should also be completed.

3. A child with chronic constipation undergoes anorectal manometry and no retrosphincteric reflex is seen after balloon dilation. The patient undergoes posterior internal sphincter myectomy for internal anal sphincter achalasia. What is the optimal amount of smooth muscle resection?

   a) 5–10 mm wide smooth muscle resection

b) 2–4 mm wide smooth muscle resection
c) Perform lateral internal sphincterotomy
d) 2 cm wide smooth muscle resection

Answer: (a.): Perform 5–10 mm wide smooth muscle resection. When performing a posterior internal sphincter myomectomy, it is important to achieve the right amount of resection as too little tissue resected leads to continued constipation and too much leads to incontinence.

## Further Reading

Arain Z, Rossi TM. Gastrointestinal bleeding in children: an overview of conditions requiring nonoperative management. Semin Pediatr Surg. 1999;8(4):172–80.

Ambartsumyan L, Rodriguez L. Bowel management in children with spina bifida. J Pediatr Rehabilit Med. 2018;11(4):293–301.

Doodnath R, Puri P. Internal anal sphincter achalasia. Semin Pediatr Surg. 2009;18(4):246–8.

Halleran DR, Sloots C, Fuller MK, Diefenbach K. Adjuncts to bowel management for fecal incontinence and constipation, the role of surgery; appendicostomy, cecostomy, neoappendicostomy, and colonic resection. Semin Pediatr Surg. 2020;29(6)

Irish MS, Caty MG, Azizkhan RG. Bleeding in children caused by gastrointestinal vascular lesions. Semin Pediatr Surg. 1999;8(4):210–3.

Lane VA, Wood RJ, Reck C, Skerritt C, Levitt MA. Rectal atresia and anal stenosis: the difference in the operative technique for these two distinct congenital anorectal malformations. Techn Coloproctol. 2016;20(4):249–54.

Stites T, Lund DP. Common anorectal problems. Semin Pediatr Surg. 2006;16(1):71–8.

Wood RJ, Levitt MA. Anorectal malformations. Clin Colon Rectal Surg. 2018;31(2):61–70.

Zar-Kessler C, Kuo B, Cole E, Benedix A, Belkind-Gerson J. Benefit of pelvic floor physical therapy in pediatric patients with dyssynergic defecation constipation. Digest Dis (Basel, Switzerland). 2019;37(6):478–85.

# Surgical Evaluation of Skin Lesions in Childhood

**Audrey Dean and Howard Pride**

## Pigmented Lesions

**Key Points**

1. The overwhelming odds are that a pigmented lesion will be benign in childhood, the incidence of malignant melanoma being about one per million in prepubertal children, about one in 250,000 in young teenagers, and one in 50,000 in older teenagers.
2. Not everything that is pigmented is melanocytic. Smooth muscle hamartoma, Becker nevus, plexiform neurofibroma, café au lait macules, morphea, mastocytoma, and granuloma annulare might imitate a melanocytic lesion.
3. Involve pediatric dermatology or general dermatology consultation prior to large, potentially deforming procedures to assure that removal is necessary and beneficial.

A. There are several atypical appearing melanocytic pigmented lesions that are eye catching but harmless. They generally do not need removal (Fig. 1).

  i. Scalp nevi tend to have a rim of hypo or hyperpigmentation at their periphery, lending them the nickname of eclipse nevi, given their resemblance to a solar eclipse. Be aware that the scalp, axillae, and groin are histopathologic "special sites," and atypia might be reported on a biopsy specimen, even in a harmless lesion.

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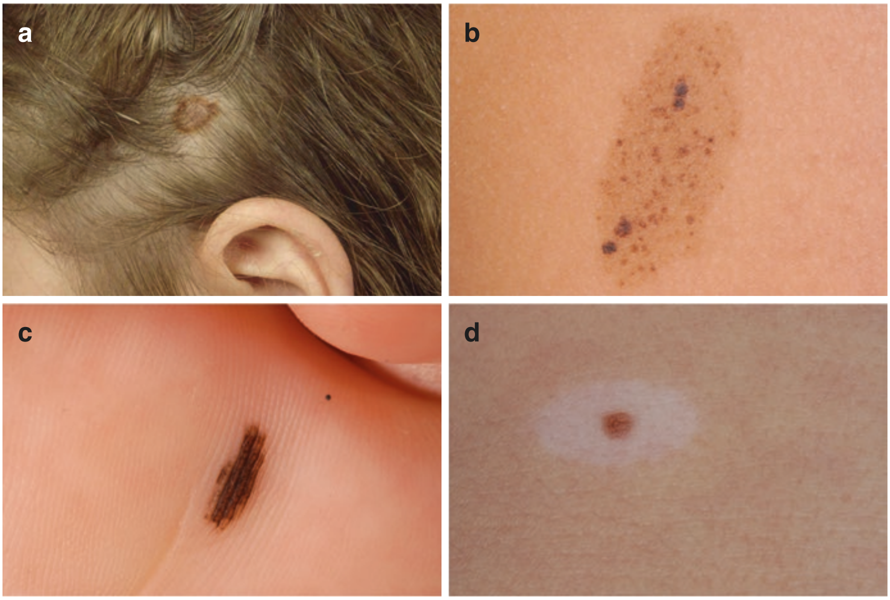


**Fig. 1** (**a**) Scalp Eclipse Nevus, (**b**) Nevus Spilus, (**c**) Acral Nevus, (**d**) Halo Nevus

ii. Nevus spilus may be congenital or acquired; the name arises from the illusion that pigment has been "spilled" across a light brown background.
iii. Acral nevi, located on the thick skin of the palms or soles, tend to have fuzzy, irregular borders and strangely reticulated, dark brown pigment. Dermoscopy can be very helpful in following these lesions and clinical follow-up in children is a better alternative to a difficult surgery with associated morbidity.
iv. Halo nevi represent an autoimmune response to nevus pigment, very akin to vitiligo. A rim of hypopigmentation is seen surrounding a central brown or red melanocytic nevus. There may be many halo nevi present in a single individual.

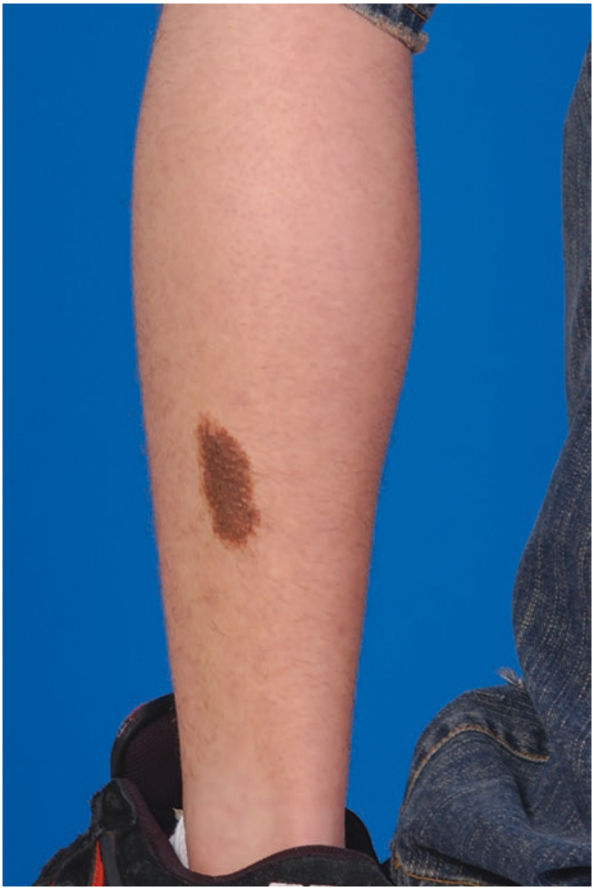
B. Congenital nevus

i. About 1% of children are born with a melanocytic nevus or will develop one in the first year of life.
ii. They tend not to follow the usual "A, B, C, D, E" rules (see below) for concerning atypical nevi and are characteristically asymmetric, irregular, and large (Fig. 2).
iii. Despite their impressive appearance, they behave in a harmless fashion and do not need to be removed for cancer prophylaxis.
iv. There is deep penetration of melanocytes into the dermis so, if considered necessary for functional or cosmetic reasons, removal needs to be done by complete elliptical excision rather than by shave.

C. Spitz nevus

i. Clinical appearance tends to take on two different morphologies: a red, dome-shaped papule resembling a pyogenic granuloma (Fig. 3a) or a deeply pigmented, mottled, or spiculated macule or plaque (Fig. 3b).

**Fig. 2** Congenital Nevus



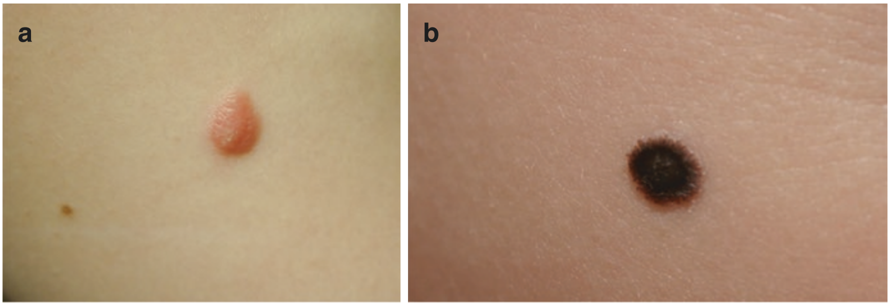


**Fig. 3** (**a**) Spitz Nevus, red, dome-shaped type. (**b**) Spitz Nevus, dark, spiculated type

ii. Pathology can mimic malignant melanoma, so it is important that these lesions be read by an experienced dermatopathologist or sent for dermatopathology consultation.
iii. Any residual lesion should be completely excised to prevent confusion with melanoma in future years. Excision is preferred over scoop shave.

iv. An atypical Spitz nevus or Spitzoid melanoma has more complex management and requires a team approach with pediatric dermatology, dermatopathology, and surgical oncology.

D. Atypical nevus and malignant melanoma

i. The A, B, C, D, E criteria are helpful in the diagnosis of an atypical nevus or malignant melanoma but may not distinguish between the two (Fig. 4).
*A*symmetry, *B*order irregularity, *C*olor irregularity, *D*iameter greater than 6 mm (size of a pencil eraser), and *E*nlarging or *E*volving.
ii. A variant of these criteria for pediatrics includes Amelanotic, Bleeding Bump, Color uniformity, De novo (new) of any Diameter, and Evolution. These criteria are sensitive but not specific for pediatric melanoma.
iii. Dermoscopy is a technique for diagnosing and following unusual, pigmented lesions.
iv. Atypical nevi of moderate cytologic atypia or greater should be completely removed by saucerization or elliptical excision and patients followed thereafter based on their number of atypical moles and family history.
v. Malignant melanoma requires complete removal with margins dictated by the histologic depth of penetration of the tumor:
In situ with 5–10 mm margins
≤1 mm with 1 cm margins
>1–2 mm with 1–2 cm margins
>2–4 mm with 2 cm margins
>4 mm with 2 cm margins
The management for children mirrors that of adults but the prognosis in children tends to be better.

E. Surgical techniques of removal

i. Parallel shave with a scalpel blade (Fig. 5a) or Personna® razor blade (Fig. 5b) is a quick and easy technique for removing symptomatic, bother-

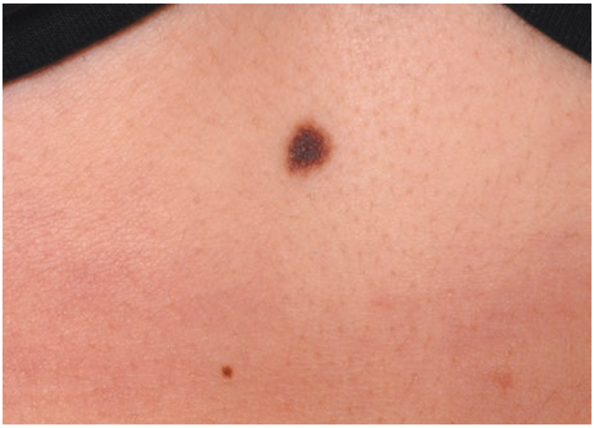


**Fig. 4** Atypical Nevus in this case but it could just as easily have been a malignant melanoma

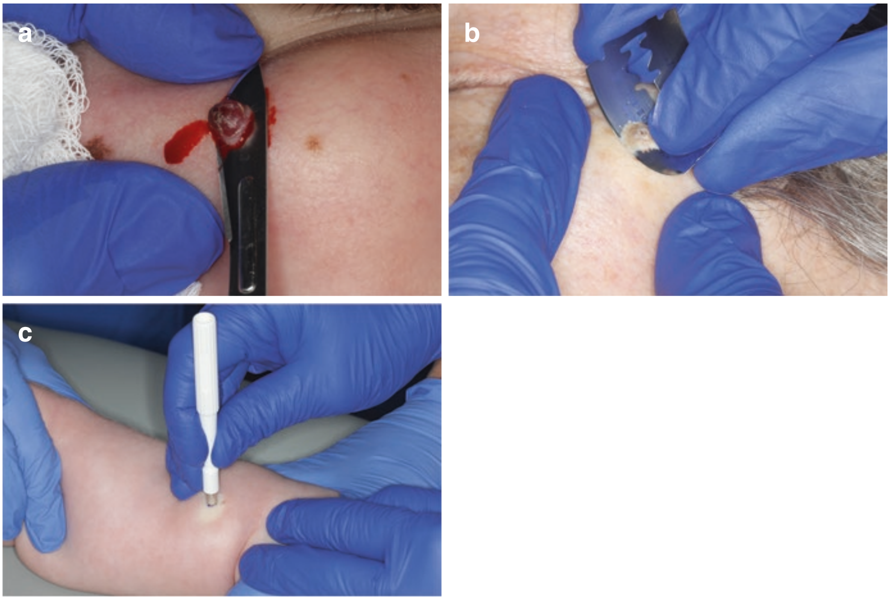


**Fig. 5** Lesion Removal Techniques (**a**) Shave with scalpel blade, (**b**) Shave with Personna® razor blade, (**c**) Punch removal

some, benign pigmented lesions. The end cosmetic result is usually excellent, but it may result in some residual melanocytes at the base.

ii. Saucerization scoop shave uses the scalpel or razor blade to scoop deeper into the dermis or even subcutaneous tissue, allowing healing to occur by second intention. It is an appropriate means for complete removal of dysplastic nevi or initial biopsy for suspected malignant melanoma.
iii. Full depth of a very small lesion can be removed with the use of a punch biopsy (Fig. 5c), but the resultant margin is slim.
iv. Elliptical excision with layered closure is the preferred method of removing congenital nevi, atypical Spitz nevi, and malignant melanoma.
v. Any removed tissue must always be sent for pathological inspection.

## Study Question

Which statement below is true of a Spitz nevus?

A. It is a childhood form of malignant melanoma with removal following the same guidelines as for melanoma.
B. It is a pre-cancerous form of malignant melanoma that needs to be removed with 5 mm margins.

C. It is pigmented but is not a melanocytic lesion and is better thought of as a mimicker of a mole.
D. It tends to have a dark, hyperpigmented ring at the periphery resembling a solar eclipse, the so-called eclipse nevus.
E. It is a benign but histologically alarming melanocytic nevus that, if removed, should be removed in its entirety to prevent future confusion with malignant melanoma.

Answer (E.) Spitz nevi in children are benign but can have histologic findings that can be misinterpreted as malignant melanoma. It is important that the pathology is interpreted by a dermatopathologist who is aware that the lesion was removed from a child. Recurrence after shave removal can be easily mistaken for malignant melanoma, so complete excision is recommended if a Spitz nevus has been only partially removed or biopsied.

## Vascular Lesions

**Key Points**

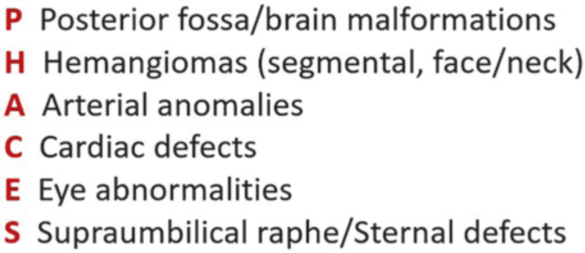
1. The most important diagnostic decision with a vascular lesion in childhood is to determine whether it is a vascular tumor or malformation, as the two entities have different associations, treatment protocols, and outcomes.
2. A vascular tumor in infancy is most commonly an infantile hemangioma, and may be treated or not, depending on its size and location.
3. A vascular malformation in childhood is most commonly a capillary malformation such as a nevus simplex (which commonly fades with time) or port wine stain (which tends to be more permanent and have associated syndromes).

A. Use the following points to help decipher between vascular tumors and vascular malformations:

    a. Vascular Tumors

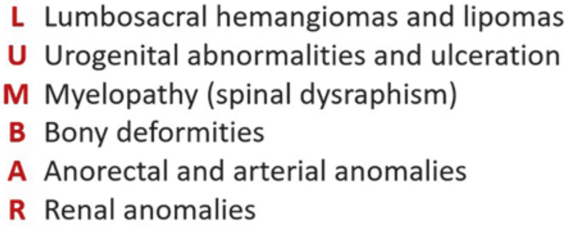
        i. Are normally not present at birth.
        ii. Have a characteristic growth period followed by involution.
        iii. May have extracutaneous involvement.
        iv. Are a neoplasm of the vasculature, therefore are mitotically active.
        v. Include infantile hemangioma, pyogenic granuloma, and kaposiform hemangioendothelioma.
        vi. May extracutaneous involvement with the following:

            1. Large, facial hemangiomas—PHACES Syndrome (Fig. 6)
            2. Large hemangiomas on lower body, especially if midline—LUMBAR Syndrome (Fig. 7)



**P** Posterior fossa/brain malformations
**H** Hemangiomas (segmental, face/neck)
**A** Arterial anomalies
**C** Cardiac defects
**E** Eye abnormalities
**S** Supraumbilical raphe/Sternal defects

**Fig. 6** PHACES Syndrome



**L** Lumbosacral hemangiomas and lipomas
**U** Urogenital abnormalities and ulceration
**M** Myelopathy (spinal dysraphism)
**B** Bony deformities
**A** Anorectal and arterial anomalies
**R** Renal anomalies

**Fig. 7** LUMBAR Syndrome

3. “Beard” distribution—airway involvement
4. Multifocal lesions—visceral, especially hepatic, hemangiomas

b. Vascular Malformations

  i. Are present at birth.
  ii. Remain unchanged or worsen with age.
  iii. Represent post-zygotic mutations with genetic mosaicism of various tumorigenic genes and may be associated with organ or limb overgrowth and syndromic associations.
  iv. Examples include capillary (such as a nevus simplex or port wine stain), venous, arterial, lymphatic, or combined malformations.

B. Vascular Tumors

a. Hemangioma

  i. Infantile hemangiomas (IH) are by far the most common type of vascular tumor, and the most common tumor of infancy (Fig. 8).

    1. Lesions can be focal or have a broader distribution, but a superficial IH has a bright red color and finely lobulated surface, regardless of size. A deeper IH will have a purple-blue color or no color change at all.
    2. Onset of an IH is within the first few weeks-months of life. They have a characteristic rapid growth phase for 2–3 months, slow growth for 5–6 more months, then a slow involution over a period of years.
    3. Treatment decisions should involve pediatric dermatology, as each lesion needs to be assessed individually and account for size, location, and other associated factors.

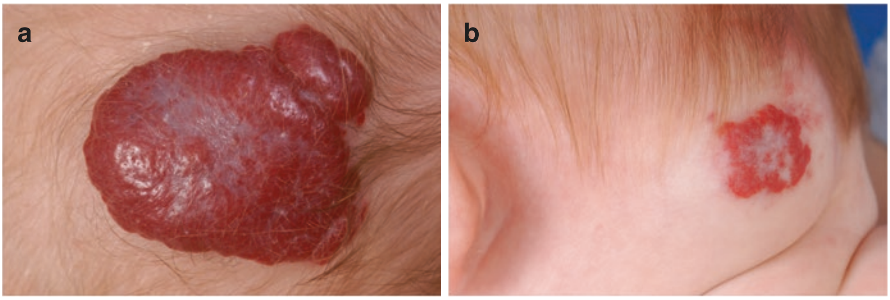


**Fig. 8** Infantile hemangiomas (**a**) Superficial types have a bright red, finely lobulated surface (**b**) Superficial and deep types have mixed appearance, with some underlying purple-blue color change

4. Many lesions involute and resolve completely, but lesions can have leftover fibrofatty, cosmetically significant residua.
5. Majority of hemangiomas can be managed with observation alone, but if an IH requires intervention, first-line therapy is done with topical timolol or systemic propranolol.
6. Surgical intervention with simple excision is considered when a lesion is unresponsive to medical treatment, or if resulting scar from excision would be preferable to leftover signs of involuted hemangioma. Surgical intervention for post-involution fibrofatty remnants or tissue redundancy can be considered.

b. Congenital hemangioma

   i. A congenital hemangioma is violaceous in color with surface telangiectasias.
   ii. These are classified as either rapidly involuting (RICH) or non-involuting (NICH).
   iii. A RICH may require surgical treatment of residual fibrofatty tissue.
   iv. Treatment of choice for a NICH is often excision.

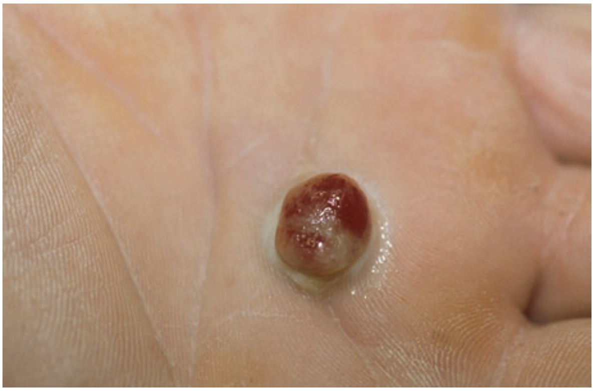
c. Pyogenic granuloma (PG)

   i. PGs rapidly appear as solitary, bright red, raised, eroded papules (Fig. 9). They are associated with trauma, pregnancy, and medications (OCPs, retinoids, indinavir, and EGFR inhibitors).
   ii. PGs are easily treated with shave removal or excision, with curettage and/or electrodessication of the base.

d. Kaposiform hemangioendothelioma, tufted angioma

   i. These are both rare vascular tumors of infancy. Each appears as a large, red-purple mass, usually involving an extremity.
   ii. These can be complicated by the Kasabach-Merritt phenomenon (KMP)—a consumptive coagulopathy leading to severe thrombocytopenia, hypofibrinogenemia, and anemia.

**Fig. 9** Pyogenic granuloma



- iii. Management should involve a team approach with pediatric hematology and dermatology.
- iv. Treatment can include surgical excision and is particularly indicated with complicating KMP.
- v. Other treatment modalities include corticosteroids, vincristine, and/or mTOR inhibitors such as sirolimus.

C. Vascular Malformations

  a. Capillary

    i. Nevus simplex—present on central forehead, eyelids (aka "Angel kiss") or on nape of neck (aka "Stork bite"); 95% of these lesions clear within the first year of life, and they do not raise concern for associated syndromes.

    ii. Port-wine stain (PWS, Fig. 10)—congenital, have more permanent appearance, and may become more violaceous and take on a raised, cobblestone texture with time.

      a. Depending on location and appearance, a PWS can raise concern for associated syndromes.

      b. Sturge–Weber syndrome (encephalotrigeminal angiomatosis) is caused by a somatic mosaic mutation in GNAQ gene. Think of this with a PWS in the V1 > V2 or V3 facial distribution. It is associated with eye abnormalities as well as meningeal and cerebral lesions. It can lead to seizures, glaucoma, intellectual disability, and focal neurologic deficits.

      c. Klippel–Trenaunay syndrome (angio-osteohypertrophy) is a syndrome involving a PWS—usually on a single extremity—which is combined with soft tissue overgrowth and lymphedema/swelling of the affected limb. This can involve limb length discrepancy leasing to orthopedic issues.

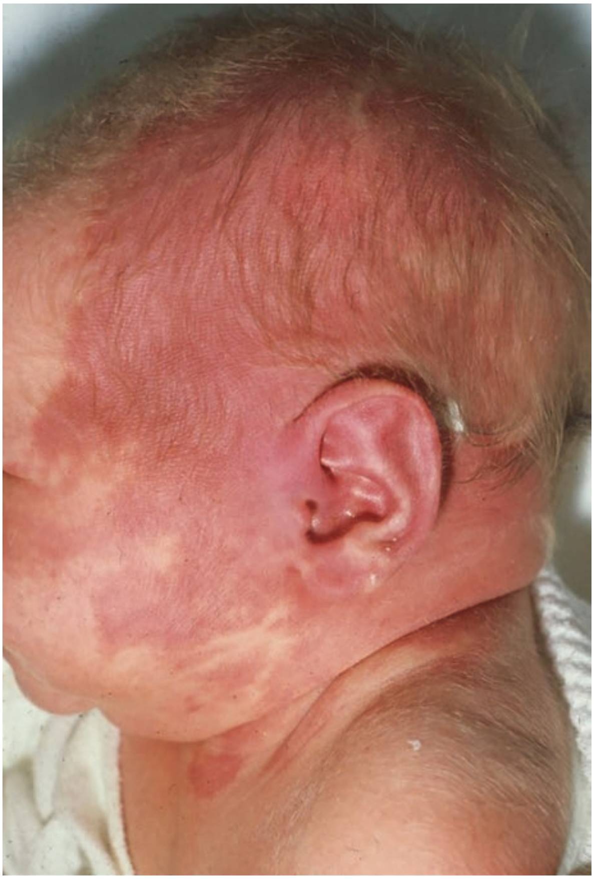


**Fig. 10** Port wine stain

iii. The most effective treatment for PWS is a pulsed-dye laser, but results vary, and most often a lightening of the lesion is achieved rather than complete clearance.

b. Venous

   i. Lesions are blue in color and present as soft, compressible nodules or plaques without warmth or pulse on palpation, or thrill on auscultation.
   ii. These can cause local distortion and discoloration, depending on location and extent of involvement.
   iii. Compression is key for conservative management, but ablative therapy can be utilized with percutaneous sclerotherapy or Nd:YAG laser ablation.

c. Lymphatic (Fig. 11)

   i. Microcystic lymphatic malformation (lymphangioma circumscriptum, verrucous lymphangioma).

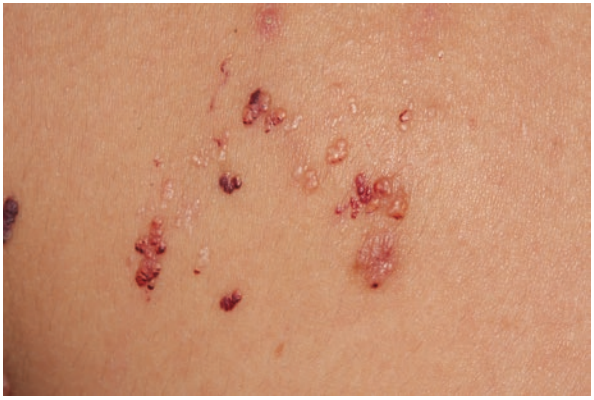


**Fig. 11** Lymphatic malformation

1. These present in one anatomic location with clusters of papulovesicles of clear-red color. They are given the descriptive nickname "frog spawn" as they resemble a frog's bumpy back.
2. Lesions can be treated/removed with excision or destructive measures ($CO_2$ or pulsed-dye laser) if small and localized and/or in a cosmetically sensitive area. Recurrence is common.

ii. Macrocystic lymphatic malformation (cystic hygroma)

1. This is associated with fetal aneuploidy such as Turner syndrome or Down syndrome.
2. This presents as a large, soft mass, most commonly on neck or axilla.
3. This can have infection or hemorrhage as complications, in addition to associated chromosomal abnormalities.

## Study Question

A newborn infant has a maroon-colored patch over her left frontal scalp, left upper eyelid, and left forehead. Is this birthmark associated with other issues, and if so, should she be seen by any other specialists at this time?

A. No, this lesion resolves with time and is not associated with any syndrome.
B. No, this lesion is not associated with any syndrome, but it will grow and spread before eventually involuting.
C. Yes, this lesion can be associated with posterior fossa abnormalities, cardiac and arterial malformations, eye issues, and midline truncal defects.
D. Yes, this lesion can be associated with Sturge-Weber syndrome. Neurology and Ophthalmology should be consulted for further evaluation.

Correct Answer: (D.)

The lesion described is a port-wine stain (PWS), a capillary malformation. The PWS is described in the V1 distribution, which can be associated with Sturge–Weber syndrome. Neurology should help evaluate for leptomeningeal or cerebral involvement, and ophthalmology can help determine whether there is associated eye pathology. Answer A describes a nevus simplex, which is present on the mid-forehead, occasionally eyelids, and/or nape of the neck. Answers B and C represent infantile hemangiomas.

## Common Benign Cutaneous Lesions of Childhood

**Key Points**

1. Most cutaneous lesions in children are benign and can be clinically observed, but surgical removal of some lesions may be considered based on location or expected course. Consultation with dermatology is appropriate to assist with a treatment plan.
2. Midline cutaneous lesions, particularly on the face and lumbosacral area, warrant additional imaging.

1. Juvenile xanthogranuloma (JXG)
   a. A JXG appears as a round, dome-shaped, red-brown papule that becomes more yellow with time (Fig. 12). They are usually solitary and appear in the first year of life.
   b. When there are numerous JXGs (3 or more) present and in patients less than 2 years old, consider evaluation for ocular involvement.

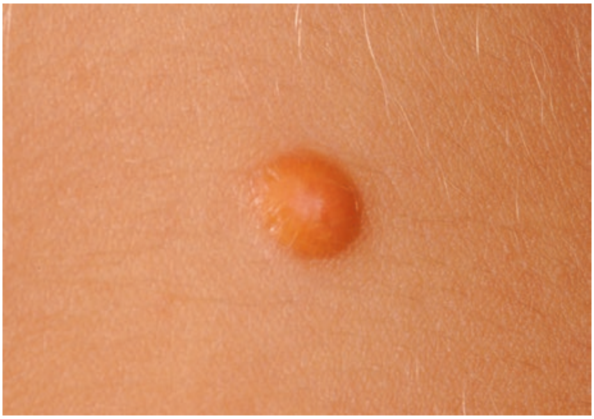


**Fig. 12** Juvenile xanthogranuloma JXG

   c. Surgical removal of JXGs is typically not recommended given their usual spontaneous involution. Rarely, giant JXGs occur, and these may require surgical intervention. This is particularly the case when removal would avoid ulceration or difficult wound management or if fibrofatty remnants of a lesion, post-involution, would leave more disfigurement than a scar from early primary closure.

2. Nevus sebaceous
   a. This is a common, congenital epidermal hamartoma, a subtype of epidermal nevus.
   b. A nevus sebaceous is present at birth as pink-tan, hairless, bumpy plaque on head/neck (Fig. 13) but undergoes changes at puberty to be more yellow in color and verrucous in texture.
   c. These can develop other benign, or more rarely malignant, neoplasms within it over time (most commonly benign trichoblastomas). The lesion can be unsightly and bothersome as it undergoes changes during and after puberty, so they are often removed.
   d. Treatment of choice is elliptical excision.
3. Midline nasal lesions
   a. The differential diagnosis of midline nasal lesions includes dermoid cyst, encephalocele, nasal glioma, hemangioma, epidermoid cyst.
   b. Some form of imaging study must be performed to rule out central nervous system connection prior to any surgical procedure.
4. Dermoid cyst
   a. This is seen in infants as a firm, deep-seated nodule that is regular in shape. They most commonly present on lateral eyebrow but can also present on midline nose, over anterior fontanelle, or on submental area.

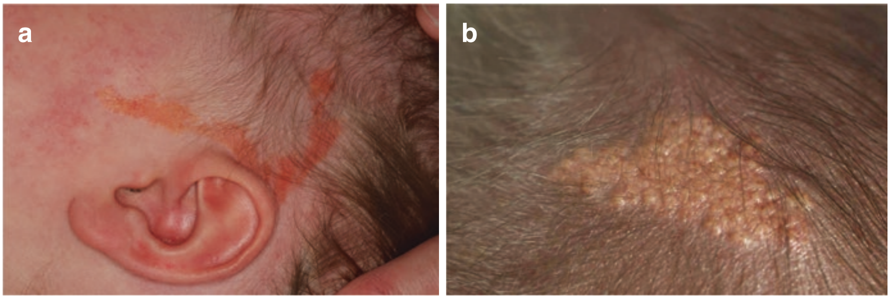
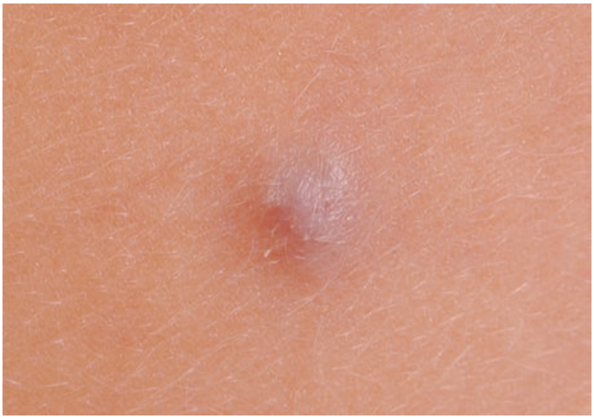


**Fig. 13** Nevus sebaceous. (**a**) Appearance at birth as pink-tan, hairless plaque (**b**) Appearance of similar lesion as it was changing during adolescence into yellow verrucous plaque

5. Encephalocele
    a. This involves intracranial structure(s) herniating through a defect in the skull.
    b. This appears as a soft nodule or mass that transilluminates and increases in size with crying or Valsalva maneuver. It may have a translucent, glistening surface.
    c. Most commonly this presents on occiput or vertex scalp, but it can also present midline on nasal glabella (in which case is called a frontal encephalocele).
6. Nasal glioma
    a. This is analogous to frontal encephalocele, but it does not have an intact intracranial connection.
    b. This is firm and does not have a size increase with crying or Valsalva maneuver.
7. Pilomatricoma
    a. This is seen in young children on the face or scalp most commonly. It can have a whitish color if superficial or a bluish hue if deep (Fig. 14).
    b. This has an irregular shape and feels, akin to "meteorites" on palpation. They are described to display two clinical signs, which can be particularly helpful for deeper lesions:
        i. Tent sign—when overlying skin is stretched, multiple angles and points seen.
        ii. Teeter-totter sign—when one side of lesion is pushed down, the other side pops up to skin surface.
    c. The treatment of choice for enlarging or cosmetically bothersome lesions is surgical removal, which often can be done with a small incision and dissection of the intact tumor. After removal, subcutaneous tissue should be lavaged with saline to remove any possible particulate calcifications.

**Fig. 14** Pilomatricoma



8. Midline sacral lesions
    a. Lesions on the midline sacral area should raise concern for occult spinal dysraphism (incomplete fusion of posterior midline embryonic structures).
    b. A wide variety of cutaneous lesions have been associated with increased risk of underlying pathology, including sacral dimple, sinus tract, aplasia cutis, hairy patch, lipoma, hemangioma or vascular malformation, pigmented lesion, and others.
    c. The presence of two or more of the cutaneous signs on midline sacral area raises concern for spinal dysraphism, in which case imaging should be done.
    d. In patients 3–4 months in age, imaging can be done with ultrasound, but MRI provides the best, most accurate answer.
    e. Biopsy or surgical removal of lesions suspicious for underlying pathology should not be performed without diagnostic imaging and referral to neurosurgery, as introduction of infection into spinal canal can occur.
9. Lipoma
    a. While quite common in adults, lipomas are not frequently seen in children.
    b. A lipoma in lumbosacral area is of particular importance given sequalae associated with midline sacral lesions (see above).
    c. A lipoma presents as a firm, rubbery, mobile subcutaneous nodule. They are most often painless but occasionally tender to palpation, especially if it is an angiolipoma.
    d. If lipoma is particularly symptomatic or is progressively enlarging, it is best removed when small. This can be done with minimal incision or "squeeze" technique utilizing extraction through a hole created by small punch biopsy or small scalpel incision. It is important to remove the entire fibrous capsule to avoid recurrence.
10. Digital fibroma (infantile digital fibromatosis, aka Reye tumor)
    a. This is a firm, red-brown mass that may be present at birth or within the first year of life on fingers or toes, although characteristically they spare the great toe and thumb.
    b. Most lesions spontaneously regress, therefore nonsurgical observation can be appropriate, but intervention is warranted if there is functional impairment.
    c. Recurrence rate after excision is high—50–75% reported. Mohs micrographic surgery has been utilized more recently with no recurrence in a few reported cases.
11. Dermatofibroma
    a. This is a firm, small nodule, most commonly on extremities; it may demonstrate the "dimple sign" with central puckering of the lesion when pinched laterally.

  b. These are common and believed to develop after minor trauma such as insect bites.
  c. Simple excision is treatment of choice for symptomatic or cosmetically unacceptable lesions, but most often they are best left alone.

12. Accessory tragus, preauricular sinus—lesions secondary to abnormalities of the first branchial cleft and arch.

  a. Accessory tragus

    i. These have a similar appearance to a skin tag (Fig. 15).
    ii. These are found along the preauricular area or neck—along the course of migration taken by branchial arches during development (along imaginary line drawn between tragus and corner of mouth or on neck along anterior edge of sternocleidomastoid muscle).
    iii. These can be single, or multiple as well as unilateral or bilateral.
    iv. Excision of an accessory tragus should include careful removal of any underlying cartilage present, which can extend deeply.

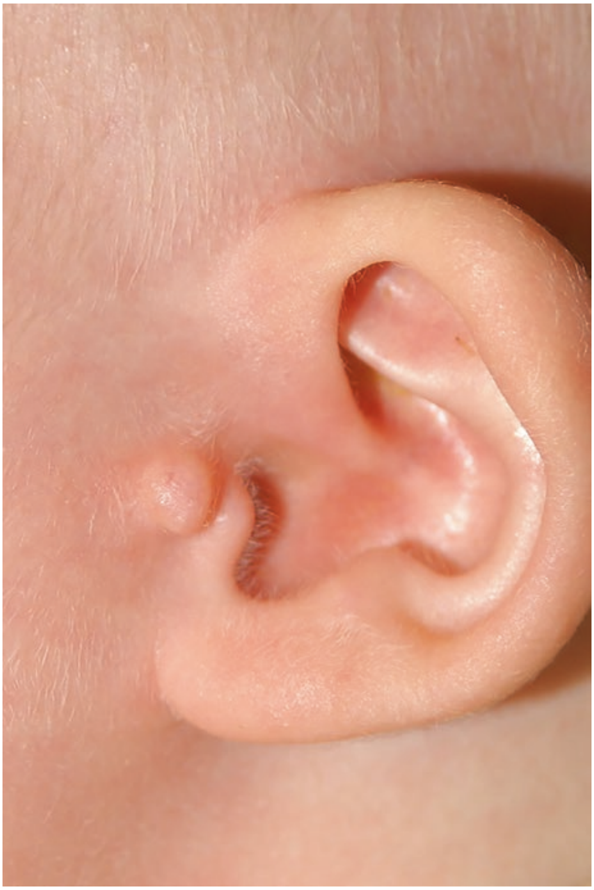


**Fig. 15** Accessory tragus

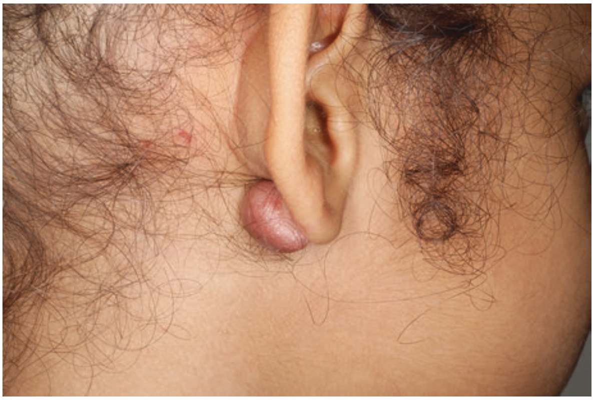


**Fig. 16** Keloid

b. Preauricular sinus
    i. This can present as enlarging mass or draining sinus in anterior or posterior auricular skin or anterior neck.
    ii. This is less common than accessory tragus. It is anomaly of first branchial cleft.
    iii. This can occur alone or with hearing loss.
    iv. Complete surgical excision can prevent complications later in life, and is best done by an otolaryngologist.

13. Keloid
    a. Dense, fibrous nodule (Fig. 16) which develops at the site of previously traumatized skin (surgical site, piercing, burn, etc.).
    b. Management can be very difficult; should begin with a series of intralesional steroid injections and progress to excision only with careful consideration of postoperative plan for avoiding recurrence. Other nonsurgical interventions (employed in place of surgery or perioperatively) include cryotherapy, laser therapy, radiation, and injection with 5-fluorouracil or interferon.

## Study Question

A 9-month-old child presents with a firm, 6 mm round subcutaneous nodule on the midline nasal root. What should be done before any surgical treatment is considered?

A. Ophthalmology consult
B. Neuroimaging
C. Children and Youth Services
D. Pulsed dye laser treatment

Correct Answer: (B.)

Midline nasal lesions can have a connection to the central nervous system. This should be fully evaluated with appropriate imaging prior to any surgical treatment. Answer A is not correct, as eye involvement would not be suspected in this case. Answer C is incorrect, as this lesion is not suspicious for non-accidental trauma. Answer D is incorrect because pulsed dye laser treatment would not be appropriate for this lesion.

## Malignant Cutaneous Lesions of Childhood

**Key Points**

1. Malignant cutaneous lesions require a collaborative approach with pediatric oncology, surgical oncology, Mohs surgery, and pediatric dermatology.
2. The development of basal cell carcinoma (BCC) or squamous cell carcinoma (SCC) in childhood should prompt a search for other features of an underlying cancer-prone genetic syndrome.
3. Controlled margins with Mohs micrographic surgery rather than wide local excision are preferred for removal of dermatofibrosarcoma protuberans (DFSP), and certain squamous cell carcinomas and basal cell carcinomas.

A. Dermatofibrosarcoma Protuberans (DFSP)

  i. DFSP is a rare, locally aggressive fibrous tumor that usually occurs on the trunk and proximal upper extremities but can also be on the head and neck. Incidence is about one per million pediatric population.
  ii. Lesions are erythematous to bluish nodules and plaques that grow slowly and seldom metastasize. Biopsy confirms the diagnosis. Local recurrence after narrow-margin excision is the norm.
  iii. Wide local excision with 2–4 cm margins has traditionally been recommended, but Mohs micrographic surgery can achieve tissue sparing with fewer recurrences.
  iv. Plastic Surgery reconstruction may be needed after removal.
  v. Lifelong follow-up is recommended.

B. Basal Cell Carcinoma (BCC)

  i. Nodular BCC typically presents as a translucent (pearly) papule or nodule with central dell or ulceration and prominent arborizing telangiectasia (Fig. 17), but BCC can also have superficial, pigmented, and morpheaform morphology. Shave or punch biopsy confirms the diagnosis.

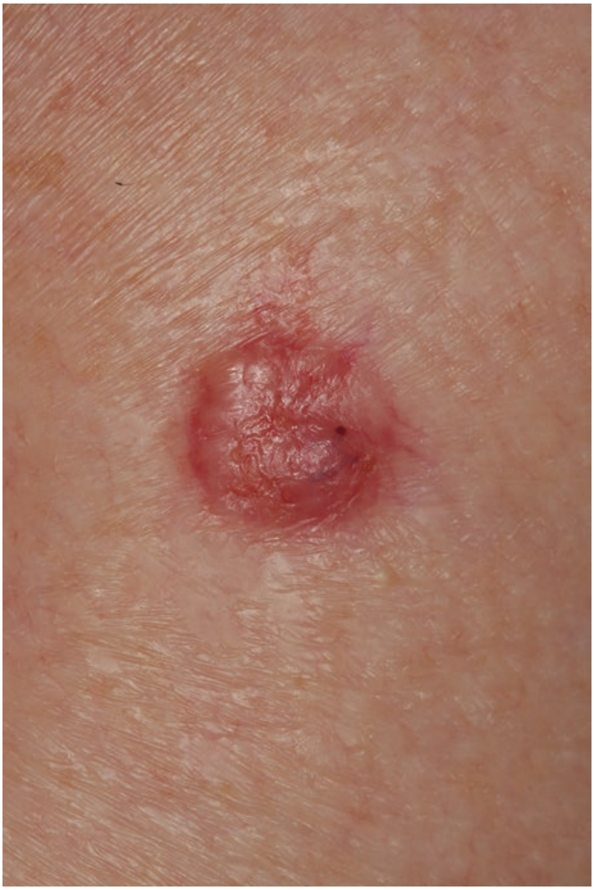


**Fig. 17** Basal cell carcinoma

ii. Thirty-six percent of pediatric BCC patients have underlying basal cell nevus syndrome (Gorlin syndrome), an autosomal dominant genetic disorder characterized by craniofacial abnormalities, palmar pits, jaw cysts, bifid ribs, and intracranial calcifications. Other genetic abnormalities that impart an increased risk include xeroderma pigmentosa, Bazex syndrome, and Rombo syndrome. Twenty percent of children with BCC have had preceding radiation or chemotherapy.
iii. Scoop shave excision with 5 mm margins followed by curettage and/or electrodesiccation is suitable for small lesions on the trunk and extremities. Mohs micrographic surgery is best for large lesions or those on the head and neck.
iv. Topical imiquimod, tazarotene, and 5-fluorouracil could be considered more child-friendly alternatives to surgery.

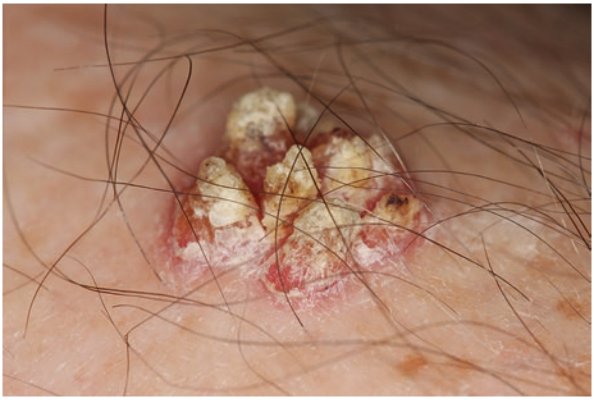


**Fig. 18** Squamous cell carcinoma

C. Squamous Cell Carcinoma (SCC)

   i. SCC presents as a scaling, indurated papule or plaque, sometimes with a very hyperkeratotic surface, most often in sun-exposed skin (Fig. 18).
   ii. SCC can occur in areas of chronic trauma, thermal burns, radiation fields, and the chronic wounds of individuals with the most severe forms of epidermolysis bullosa. It is difficult to diagnose in these settings and behaves in a more aggressive fashion with frequent recurrence and metastasis.
   iii. Thirty percent of pediatric patients with SCC have had at least 6 months of immunosuppression. Twenty-eight percent of pediatric patients with SCC have xeroderma pigmentosum, which imparts a 10,000 times increased risk.
   iv. Treatment is the same as for BCC above, but involvement of a tumor board and surgical oncologist is advised for complicated lesions such as those found in burn scars or in the setting of epidermolysis bullosa.
   v. Photodynamic therapy (topical aminolaevulinic acid followed by irradiation with blue or red light) is an additional non-surgical option for SCC.

## Study Question

A child with a basal cell carcinoma has a very high probability of also having:

A. Multiple café au lait macules and neurofibromas indicative of neurofibromatosis.
B. Severe atopic dermatitis treated with narrowband ultraviolet B (nbUVB) light therapy.
C. Palmar pits, jaw cysts, and intracranial calcifications indicative of Gorlin syndrome.
D. White macules, connective tissue nevi, and facial angiofibromas indicative of tuberous sclerosis.
E. Web neck and short stature indicative of Turner syndrome.

Answer (C.) Gorlin syndrome or basal cell nevus syndrome accounts for about 36% of all children with a basal cell carcinoma. It is an autosomal dominant genetic disorder characterized by craniofacial abnormalities, palmar pits, jaw cysts, bifid ribs, and intracranial calcifications. None of the other answers carry an increased risk of basal cell carcinoma.

## Further Reading

Campbell LB, Petrick MG. Mohs micrographic surgery for a problematic infantile digital fibroma. Dermatol Surg. 2007;33(3):385–7. https://doi.org/10.1111/j.1524-4725.2007.33080.x.

Moustafa D, Neale H, Hawryluk EB. Trends in pediatric skin cancer. Curr Opin Pediatr. 2020;32:516–23.

Rubio GA, et al. Incidence and outcomes of dermatofibrosarcoma protuberans in the US pediatric population. J Craniofac Surg. 2017;28:182–4.

Schachner L, Hansen R, Krafchik B, Lucky A, Paller AS, Rogers M, Torello A. Pediatric dermatology. 4th ed. Elsevier; 2010.

Wong JR, Harris JK, Rodriquez-Galindo C, et al. Incidence of childhood and adolescent melanoma in the United States; 1973-2009. *Pediatrics*. 2013;131:846–54.

Yang C, Gru AA, Dehner LP. Common and not so common melanocytic lesions in children and adolescents. *Pediatr Dev Pathol*. 2018;21:252–70.

# Pediatric Gastrointestinal Bleeding

**Biren Desai and Dean R. Focht**

**Key Points**

1) Hematemesis is indicative of an upper GI bleed, occurring proximal to the ligament of Treitz, while hematochezia usually indicates a lower GI bleed, distal to the ligament of Treitz.
2) A patient's age, along with a detailed history, can be used to guide diagnostic evaluation and treatment.
3) Severe GI bleeding can quickly result in hemodynamic compromise. Initial management of patients with severe GI bleeding should focus on stabilization and include quick intravascular access for fluid resuscitation.

## Introduction

GI bleeding in the pediatric population encompasses many differentials as bleeding can occur anywhere from the oropharynx to the rectum. Unlike their adult counterparts, congenital anomalies must also be considered in the differential of pediatric GI bleeding. This chapter will first focus on basic commonalities in presentation, evaluation, and laboratory findings when comparing upper and lower GI bleeding. The content will then discuss the differential diagnosis of GI bleeding both proximal and distal to the ligament of Treitz.

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## Presentation

1) Hematemesis is indicative of bleeding proximal to the ligament of Treitz and can be described as either frankly bloody or coffee ground in nature. Upper GI bleeding can also present as melena.
2) Hematochezia usually indicates bleeding distal to the ligament of Treitz, however, brisk upper GI bleeding along with a rapid intestinal transit time can occasionally result in hematochezia.
3) Mild GI bleeding may be clinically asymptomatic or present with iron-deficient anemia.
4) Severe GI bleeding can result in hemodynamic compromise, including tachycardia, hypotension, or shock.

## Evaluation

1) Key Topics in the History:
    a) Determine if the blood loss is acute or chronic.
    b) Estimate the amount of blood loss.
    c) Is the patient having coffee ground or frankly bloody emesis, hematochezia, or melena?
    d) Is the patient taking any medications? Particular attention should be given to blood thinners, NSAIDs, and medications known to cause pill esophagitis.
    e) Is there associated abdominal pain? If yes, where is the pain located and when did it start?
    f) Are there any other associated symptoms related to the GI bleed, including fevers, diarrhea, constipation, weight loss, or mental status changes?
    g) Are there any other underlying medical problems that may predispose the patient to a GI bleed, such as cirrhosis or underlying liver disease?
    h) Does the patient have a known bleeding diathesis or have there been prior episodes of GI bleeding?
    i) Make note if there have been any known contacts with individuals having similar symptoms.
2) Physical Examination Findings:
    a) A patient's vital signs can range from no derangements to those consistent with hemorrhagic shock.
    b) Assess the patient's mental status.
    c) Pay careful attention to the HEENT examination to evaluate for epistaxis, petechiae, or evidence of previous bleeding in the oropharynx.
    d) A thorough abdominal exam should assess for signs of pain or distention.
    e) An evaluation for signs of liver disease may reveal hepatomegaly, jaundice, scleral icterus, spider angiomas, or caput medusae.
    f) Evaluate capillary refill and look for signs of pallor.

g) Perform a complete skin exam looking for hemangiomas.
h) A rectal exam can evaluate for fissures, perianal abscesses, fistula, and hemorrhoids.
i) Verify if blood is present in the stool at the time of rectal exam as certain foods and medicines can mimic blood, including red food dyes, beets, red licorice, Kool-Aid, blueberries, iron supplements, cefdinir, and bismuth-containing medicines. False-negative hemoccult results can occur if patients are taking high doses of vitamin C.

## Laboratory/Radiology Findings

1) The following STAT labs should be considered as part of a patient's initial workup: Complete Blood Count with differential (CBC/Diff), hepatic function panel, renal function panel, prothrombin time (PT), partial thromboplastin time (PTT), international normalized ratio (INR), urinalysis, and blood type with crossmatch.
2) Further laboratory testing, which may be considered based on the patient's presenting symptoms and differential diagnosis, may include iron studies, *H. pylori* stool antigen, stool culture, stool pathogen panel PCR, and *Clostridium difficile*.
3) Hemoglobin levels may initially be normal, even in patients with significant GI bleeding, as it may take several hours to days for a patient's hemoglobin level to fully equilibrate.
4) Patients with cirrhosis may have thrombocytopenia because of splenic sequestration of platelets.
5) Elevated liver enzymes may be seen in patients with underlying liver disease.
6) An elevation in a patient's BUN may occur as blood is degraded within the lumen of the small intestine and large bowel.
7) Coagulation derangements may signify synthetic liver dysfunction that requires correction to aid in the management of GI bleeding.
8) Slow indolent bleeds can often cause iron deficiency anemia as evidenced by low ferritin or iron levels and high total iron-binding capacity.
9) A nasogastric tube with gastric lavage can evaluate for an upper GI source of bleeding.
10) Angiography can be used to identify sources of bleeding that are >0.5 mL/min. A tagged RBC scan can identify sources of bleeding that are >0.1 mL/min.

## **Upper GI Bleed Differential Diagnosis** (Table 1)

1) Epistaxis—Swallowed blood from brisk epistaxis can be mistaken as an upper GI bleed. Blood often acts as an irritant to the gastric mucosa, which can result in emesis. If enough blood traverses the GI tract, epistaxis can also lead to melena.

**Table 1** Differential diagnosis for Upper GI bleeding based on age

| Neonate | Infant/Toddler | Young children | Adolescents |
|---|---|---|---|
| Epistaxis | Epistaxis | Epistaxis | Epistaxis |
| Swallowed maternal blood | Mallory–Weiss tears | Mallory–Weiss tears | Mallory–Weiss tears |
| Stress gastropathy | Esophagitis | Esophagitis | Esophagitis |
| | Foreign body ingestion | Foreign body ingestion | Aortoenteric fistula |
| | Stress gastropathy | Esophageal varices | Esophageal varices |
| | Caustic ingestion | Caustic ingestion | Caustic ingestion |
| | Zollinger–Ellison Syndrome | Stress gastropathy | Stress gastropathy |
| | | Peptic ulcer disease | Peptic ulcer disease |
| | | Zollinger–Ellison Syndrome | Zollinger–Ellison Syndrome |

2) Mallory–Weiss tears—These are caused by repeated episodes of forceful retching or vomiting. Most Mallory–Weiss tears are located in the distal esophagus near the GE junction.
3) Caustic ingestion–The average household contains many products that can cause severe esophagitis or gastritis if consumed. Ingestion of acidic or alkali solutions can result in inflammation severe enough to cause GI bleeding.
4) Foreign body ingestion–Children often ingest foreign bodies, which can cause local mucosal damage as the object passes through the esophagus and into the stomach. Esophageal button batteries are one example of a foreign body that requires emergent removal.
5) Esophageal varices–Varices are most often caused by portal hypertension in the setting of hepatic cirrhosis. Biliary atresia and Alagille Syndrome are the two most common causes of biliary obstruction in children, which can lead to cirrhosis and development of varices. Bleeding esophageal varices are a medical emergency that carry a high rate of morbidity and mortality as patients often present with hematemesis and hypovolemic shock.
6) Esophagitis–Any irritation to the esophageal lining can cause bleeding. Immunocompromised children are at increased risk for esophagitis which can be seen in the setting of HIV, chronic steroid therapy, or in patients undergoing chemotherapy.
7) Stress gastropathy–Stress gastropathy is often seen in the critically ill child whose body is undergoing significant physiologic stress. Common underlying medical problems include sepsis, burns, head injuries, or shock. Control of the underlying pathology is important along with acid-suppressive medications.
8) Peptic ulcer disease–Ulcers are most often caused by *H. pylori* infections, NSAID use, and with high-dose steroids. GI bleeding associated with peptic ulcer disease may be indolent.
9) Zollinger–Ellison Syndrome–This is a rare condition in which a gastrin secreting tumor causes excessive stomach acid secretion, leading to multiple gastric

and duodenal ulcers. In some cases, esophageal ulcers can also be seen. The tumor can be associated with MEN 1 syndrome.

10) Aortoenteric fistula–This is a rare and life-threatening connection between the aorta and GI tract. Patients will present with brisk upper GI bleeding that can lead to hypovolemic shock and exsanguination. Prompt cardiothoracic intervention is required.

## Upper GI Bleed Management

1) Management of an upper GI bleed is based on the underlying diagnosis. An upper endoscopy can be both diagnostic and therapeutic for patients.
    a) Foreign bodies–Objects that are damaging to the mucosa of the GI tract can be removed during upper endoscopy.
    b) Peptic ulcer disease–Lesions that show stigmata of recent or active bleeding can be injected with medication, cauterized, or clipped to help prevent further bleeding.
    c) Esophageal varices–If varices are present, banding procedures can be performed to prevent bleeding.
2) For many disease processes, it is helpful to control stomach acid secretion to allow for mucosal healing. If the etiology of a GI bleed is unknown, patients should be started on an intravenous formulation of a proton pump inhibitor (PPI). This can be transitioned to an oral PPI once medically appropriate. In the setting of peptic ulcer disease, patients often require PPI therapy for several weeks to months.
3) Surgery may be required for bleeding that is difficult to control or if patients cannot be medically stabilized.

## Lower GI Bleed Differential Diagnosis (Table 2)

1) Swallowed maternal blood—This may occur during delivery or if the mom of a breast-fed infant has cracked nipples. An apt test will help distinguish maternal from fetal hemoglobin.
2) Allergic colitis—A common cause of bloody stools in both bottle and breast-fed infants. This resolves in most infants by 12–18 months of age.
3) Necrotizing enterocolitis (NEC)—NEC should be suspected in a premature newborn who develops feeding intolerance, abdominal distention, vomiting, lethargy, temperature instability, and respiratory distress. Findings on X-ray may include dilated loops of bowel or pneumatosis intestinalis. Treatment includes bowel rest and antibiotics. Surgical management may be required for

**Table 2** Differential diagnosis for lower GI bleeding based on age

| Neonate | Infant/Toddler | Young children | Adolescents |
|---|---|---|---|
| Swallowed maternal blood | Infectious colitis | Infectious colitis | Infectious colitis |
| Anal fissure | Anal fissure | Anal fissure | Anal fissure |
| Necrotizing enterocolitis | Intestinal duplication cyst | Polyps | Polyps |
| Malrotation with midgut volvulus | Very early-onset inflammatory bowel disease (VEO-IBD) | Inflammatory bowel disease (IBD) | Inflammatory bowel disease (IBD) |
| Hirschsprung disease with enterocolitis | Meckel's diverticulum | Hemolytic uremic syndrome (HUS) | Solitary rectal ulcer |
| Coagulopathy | Intussusception | Henoch–Schönlein purpura (HSP) | |
| Allergic colitis | Allergic colitis | | |

patients who develop an intestinal perforation or who fail medical management and have progression of bowel necrosis.

4) Malrotation with midgut volvulus—This should be considered in a newborn with vomiting (bilious or non-bilious) and abdominal distention. Any newborn who presents with bilious vomiting should be considered a surgical emergency until proven otherwise. A volvulus can be life threatening and requires emergent treatment. Diagnosis can be suggested by X-ray. An ultrasound can be used to make the diagnosis in a center with an experienced radiologist, but an upper GI will often best visualize the position of the duodenum.
5) Hirschsprung disease with enterocolitis—Infants with a known (or suspected) diagnosis of Hirschsprung disease who present with abdominal distention, diarrhea, and hematochezia need to be urgently evaluated for enterocolitis. A rectal tube is used for immediate decompression along with the use of appropriate antibiotics.
6) Coagulopathy—A number of coagulopathy disorders can present with GI bleeding in infants. Vitamin K deficiency should be considered in any infant who did not receive a dose of vitamin K after birth.
7) Anal fissure—Anal fissures can be a cause of hematochezia in any age group. Constipation can cause small tears in the anal mucosa leading to painful defecation and bright red blood streaked on the outside of stools.
8) Intestinal duplication cyst—These cysts may contain gastric mucosa, which can ulcerate and cause subsequent bleeding into the GI tract.
9) Very early-onset inflammatory bowel disease (VEO-IBD)—VEO-IBD is a distinct form of inflammatory bowel disease that presents in children <6 years of age. These patients may have an underlying genetic defect or immune deficiency and tend to have a more severe and difficult to control disease compared to older children who develop IBD.
10) Meckel's diverticulum—This can present in toddlers as painless rectal bleeding. Diagnosis can be made with a Meckel's scan. If suspicion is high but diag-

nostic testing is negative, or if a patient is hemodynamically unstable, exploratory laparoscopy may be necessary.

11) Intussusception—The most common presenting symptom in patients with intussusception is abdominal pain. Infants <12 months of age are more likely to present with hematochezia than older children. Diagnosis can best be made by ultrasound, although it can sometimes be detected on abdominal X-ray. An air contrast enema can be both diagnostic and therapeutic. Intussusception is usually idiopathic in infants and toddlers but more likely to have a pathologic lead point in older children.
12) Infectious colitis—If a patient presents with fever, in addition to bloody stools and tenesmus, then stool studies should be obtained looking for an infectious etiology.
13) Polyps—Polyps usually present as painless rectal bleeding in an otherwise healthy child. Most polyps in children are benign. If there is a strong family history of intestinal polyps or colon cancer, or if >3–5 polyps are detected during colonoscopy, then genetic testing may help identify a mutation to assist in diagnosis of a polyposis syndrome.
14) Inflammatory bowel disease (IBD)—IBD in children includes patients with Crohn's disease or ulcerative colitis. In children, the peak age of onset is during adolescence. Presenting symptoms vary based on disease location but may include abdominal pain, diarrhea, hematochezia, weight loss, failure to thrive, perianal fistula or abscess, arthralgia, or erythema nodosum. Patients may have anemia, thrombocytosis, elevated inflammatory markers, low iron, and elevated fecal calprotectin or lactoferrin. An upper endoscopy and colonoscopy can identify histologic evidence of IBD. Radiology studies to help localize disease and evaluate abscess or fistula formation include an upper GI with small bowel follow-through, abdominal computed tomography (CT), or magnetic resonance enterography (MRE). An exam under anesthesia can also be used to evaluate suspected perianal fistula. Complications which may require surgical intervention include abscess or fistula formation, intestinal obstruction, perforation, intractable bleeding, toxic megacolon, or patients who are refractory to medical management.
15) Hemolytic uremic syndrome (HUS)—HUS is defined by the triad of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury. It develops as a complication of an infection with a Shiga toxin-producing *Escherichia coli* (STEC), such as *E. coli* O157:H7. Patients usually present with bloody diarrhea, and diagnosis is based on stool culture results.
16) Henoch–Schönlein purpura (HSP)—HSP is a self-limited systemic vasculitis that occurs in childhood. Clinical symptoms may vary in their presentation but include palpable purpura, abdominal pain, arthralgia, or arthritis, and renal disease. Many patients will have hemoccult-positive stools. While there are no diagnostic laboratory tests, patients should have a normal platelet count and coagulation profile. A urinalysis and creatinine should be obtained, looking for renal involvement. An abdominal ultrasound may be helpful in patients with

severe abdominal pain as edema and purpuric lesions within the intestines can serve as a lead point for intestinal intussusception.

17) Solitary rectal ulcer—This is an uncommon cause of rectal bleeding in children and adolescents, which may be associated with rectal prolapse or straining during defecation. Diagnosis may be made incidentally during colonoscopy. Treatment can include stool softeners, stool bulking agents, and biofeedback to treat dysfunctional defecation.

## Lower GI Bleed Management

- Initial management of GI bleeding involves patient stabilization. In cases of severe bleeding, quick IV access and fluid resuscitation are very important.
  - Closely monitor vital signs.
  - Administer isotonic crystalloid solutions or transfuse packed red blood cells as needed.
- Patients with severe GI bleeding who are unable to be medically stabilized may require urgent surgical intervention.

## Study Questions

Question 1: A 1-week old, former term infant, presents to the emergency department with a 5-h history of irritability, hematochezia, and bilious emesis. On exam the patient has a temperature of 101.3, heart rate of 171, and respiratory rate of 60. Abdomen is distended, tender to palpation, with hypoactive bowel sounds. Which of the following is most appropriate?

a) Emergency exploratory surgery
b) Obtain a CT scan with IV and oral contrast
c) Perform an emergent upper endoscopy
d) Obtain an upper GI series

Correct answer: (A) Bilious emesis in any infant is abnormal and should raise suspicion for malrotation. The above patient with fever, bilious emesis, distended abdomen, hypoactive bowel sounds, and hematochezia suggests a malrotation complicated by volvulus and perforation. This is a potentially life-threatening condition that requires rapid resuscitation, antibiotics, and prompt surgical exploration. No further testing is needed.

Question 2: A 16-year-old female with a recent right knee injury presents to the emergency department with 5 hours of coffee ground emesis. She appears confused; thus, history is provided by her family who states that she has been complaining of a dull abdominal ache for the last week. On examination, she is afebrile, heart rate is 165, and respirations are 22. There is no history of melena, hematochezia, or abdominal trauma. The only medication she has been taking is Naproxen. All the following diagnostic and therapeutic interventions should immediately be taken EXCEPT:

a) Obtain a CBC/Diff, hepatic function panel, renal function panel, coagulation studies, and type/crossmatch
b) Start an IV proton pump inhibitor
c) Proceed with an upper endoscopy for further evaluation
d) Administer a crystalloid intravenous fluid bolus

Correct answer: (C) This patient is experiencing an upper GI bleed. Prompt IV fluid resuscitation should begin as this patient is demonstrating altered mental status changes along with tachycardia. Obtaining basic lab work is important to help guide treatment. Until the etiology of the upper GI bleed is known, this patient should be provided an IV proton pump inhibitor medication as peptic ulcer disease is on the differential diagnosis. Although an upper endoscopy may eventually be needed, initial evaluation and treatment should focus on stabilization of this patient.

## Further Reading

Neidich GA, Cole SR. Gastrointestinal bleeding. Pediatr Rev. 2014;35(6):243–54.

Thomson M, et al. Acute upper gastrointestinal bleeding in childhood: development of the sheffield scoring system to predict need for endoscopic therapy. JPGN. 2015;60(5):632–6.

# Part VI
# Problems in School Age Children

# Lymphadenopathy

**James DeCou**

**Key Points**

1. The majority of enlarged palpable lymph nodes in children are benign reactive nodes due to infectious etiologies.
2. Lymph nodes enlarge due to multiple etiologies, including infection, inflammation, autoimmune disease, neoplasm, lipid storage disease, and medications.
3. Lymph node biopsy is indicated for any enlarged supraclavicular node and for any other node with suspicion for malignancy.
4. Prior to biopsy of a suspicious neck node, a compressing anterior mediastinal mass must be ruled out by history and imaging.

The lymphatic system is comprised of lymph nodes and interconnecting lymphatic vessels and ducts, as well as the spleen, tonsils, adenoids, Peyer's patches, and thymus. Combined, they concentrate and transport white blood cells throughout the body to help fight infections and other diseases. Lymphadenopathy can refer to any pathologic change in a lymph node, but generally is defined as the abnormal size, number, or firmness of lymph nodes.

1. Anatomy: There are approximately 600 lymph nodes in our bodies which mostly are concentrated in the head and neck, axillae, mediastinum, abdomen, retroperitoneum, and groin regions (see Fig. 1). Most normal nodes are less than 1 cm in diameter, however normal epitrochlear nodes are less than 0.5 cm, while normal

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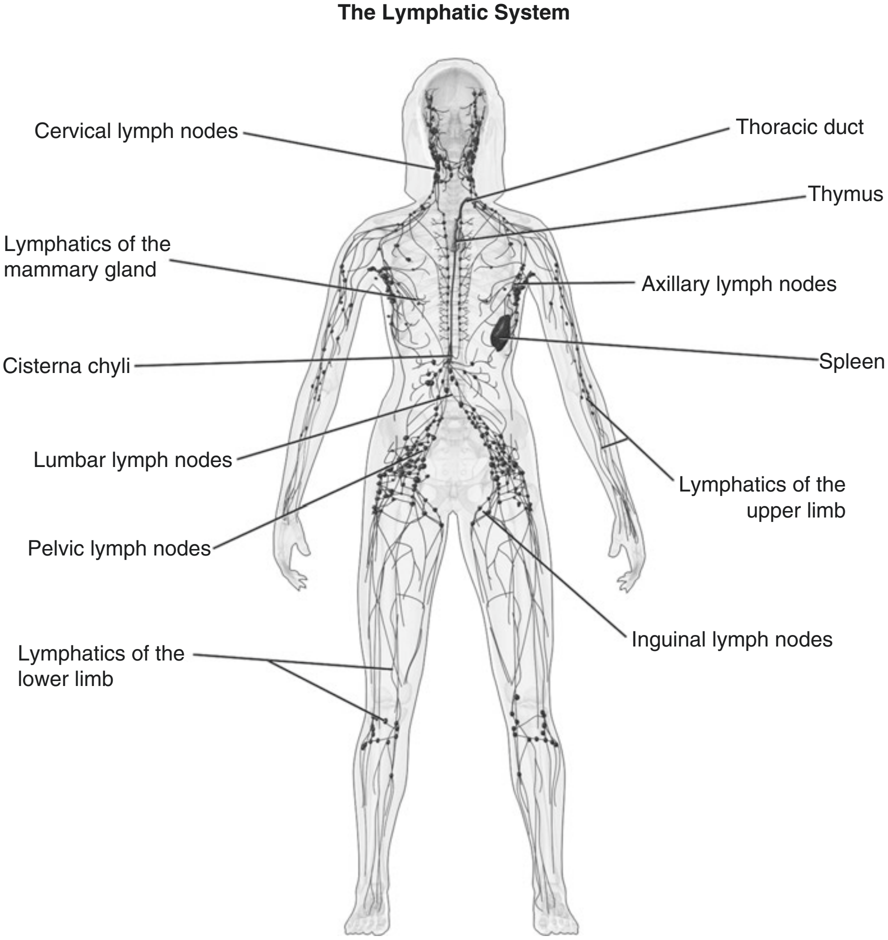
The Lymphatic System
Cervical lymph nodes
Thoracic duct
Thymus
Lymphatics of the mammary gland
Axillary lymph nodes
Spleen
Cisterna chyli
Lumbar lymph nodes
Lymphatics of the upper limb
Pelvic lymph nodes
Inguinal lymph nodes
Lymphatics of the lower limb

**Fig. 1** The lymphatic system (Source: Bruce Blausen, Blausen Medical Communications, via Creative Commons Attributions Unported 3.0 license. https://upload.wikimedia.org/wikipedia/commons/f/f4/Blausen_0623_LymphaticSystem_Female.png. Downloaded 17 Dec 2013)

inguinal nodes can be as large as 1.5 cm. Any enlargement of supraclavicular nodes is considered abnormal.

2. Epidemiology: Palpable lymph nodes are very common amongst toddlers and school-aged children. Among children 2–8 years of age, most studies report palpable nodes in about 40–60%, although some report as high as 90%. Most are benign reactive nodes due to infectious causes, with about 3% due to malignancy.
3. Pathophysiology: Lymph nodes enlarge due to infectious, inflammatory, autoimmune, neoplastic, and other etiologies. Lymphocytes work within lymph nodes to filter antigens from lymph; the extravascular fluid that travels through the lymphatic system. Lymph nodes enlarge due to several different mechanisms, most commonly a benign enlargement from processing and elimination of a

local infection. Other mechanisms include lymphadenitis and abscess due to a primary infection such as *Staphylococcus aureus*, inflammation from autoimmune disease, metastatic infiltration of neoplastic cells, localized neoplastic proliferation, and infiltration of lipids as in storage disorders.

a. Infection is the most common cause of benign, generally self-limited lymphadenopathy, and frequently is diagnosed as "reactive lymphadenopathy" on pathology exam if biopsied. Many of these can be followed expectantly without biopsy.

    i. Viral: Upper respiratory tract organisms such as rhinovirus, adenovirus, enterovirus, influenza, Epstein-Barr virus (EBV); cytomegalovirus (CMV), and HIV. SARS-CoV-2 (COVID-19) viral infection is an uncommon cause of lymph node enlargement but has been associated with cervical and mediastinal lymphadenopathy in some cases.
    ii. Bacterial: Staphylococcus, Streptococcus, *Bartonella henselae* (cat scratch disease), *Mycobacterium tuberculosis* (scrofula), Actinomyces, and nontuberculous Mycobacterium (avium-intracellulare).
    iii. Fungal: Histoplasmosis and coccidioidomycosis, both of which most commonly lead to pulmonary nodules and mediastinal or hilar lymphadenopathy.
    iv. Parasitic: Toxoplasmosis (*T. gondii*) infections frequently present with cervical lymphadenopathy.

b. Autoimmune disorders can include lymphadenopathy as one of their manifestations, with systemic lupus erythematosus and sarcoidosis being the most common of these.
c. Malignancy presenting as lymphadenopathy can be due to primary disease such as lymphoma, or it can be due to metastatic tumor spread.

    i. Lymphoma: Hodgkin and non-Hodgkin lymphoma are the most common malignancies that present as palpable peripheral lymphadenopathy. Post-transplant lymphoproliferative disorder (PTLD) is a form of lymphoma specific to solid organ and bone marrow transplant patients, and should be considered if enlarged lymph nodes are detected in these patients.
    ii. Leukemia: Lymphadenopathy can be a presenting sign of childhood leukemias.
    iii. Metastatic tumors: Pediatric malignant tumors that commonly spread to distant lymph nodes include rhabdomyosarcoma, other sarcomas, neuroblastoma, melanoma, salivary gland tumors, and thyroid carcinoma.

d. Other:

    i. Skin problems: Rashes, minor skin trauma, and animal bites.
    ii. Medications: Phenytoin is the most commonly implicated, but others include certain antibiotics (cephalosporins, sulfa drugs, penicillins), antihypertensives (hydralazine, captopril, atenolol), and anticonvulsants.

        iii. Histiocytosis: Langerhans cell histiocytosis can present with peripheral lymphadenopathy. Rosai–Dorfman disease is a histiocytic infiltration of lymph nodes that can cause massive lymphadenopathy, especially cervical.
        iv. Lipid storage diseases: Lipid infiltrates can cause lymphadenopathy in Gaucher disease and Niemann–Pick disease.

4. Clinical Evaluation:
    a. History:
        i. Recent illness, infections, bites, trauma, travel, exposure to tuberculosis or new animals.
        ii. Constitutional symptoms: fever, unintentional weight loss, night sweats, fatigue.
        iii. History of dyspnea, especially when lying flat. This is a symptom concerning for a compressing anterior mediastinal mass.
        iv. Family history of any malignancies or inherited diseases.
    b. Physical exam:
        i. Full body examination for local or generalized enlargement of lymph nodes, hepatosplenomegaly, skin rashes, nevi, scratches, or other trauma.
            1. Small nodes are palpable in most children.
            2. Enlarged nodes may be tender or nontender, mobile or fixed, discrete or matted, and soft, rubbery, or firm. If suppurative, they may be erythematous, fluctuant, or draining.
        ii. Examination of the skin surrounding enlarged nodes may show signs of infection, rash, or trauma.
    c. Studies—labs and imaging are needed only if there is suspicion for an etiology that requires further treatment (concern for malignancy, TB, or suppurative nodes). Most cases of reactive lymphadenopathy are established through a complete history and physical examination and can be safely observed with close clinical follow-up.
        i. Labs:
            1. CBC with differential if suspicious of leukemia, lymphoma, EBV, CMV, or toxoplasmosis.
            2. LDH if suspicious of leukemia or lymphoma.
            3. TB skin test.
            4. EBV, CMV, *Bartonella henselae* (cat scratch), toxoplasmosis, HIV and COVID-19 testing, as indicated.
        ii. Imaging:
            1. CXR: if suspicious of TB or hilar/mediastinal adenopathy, and if supraclavicular lymphadenopathy is present.

    2. US: is useful when there is a question of whether or not a lump is in fact an enlarged lymph node vs. dermoid vs. cyst vs. abscess vs. other.
    3. CT: can detect deep lymph nodes; may detect a source of malignancy if this is suspected; can further evaluate a mediastinal mass and possible airway compression.
    4. PET: often ordered by Hematology-Oncology if malignancy is suspected.

5. Treatment:
    a. Expectant observation: If the suspicion of malignancy or major illness is low, the majority of enlarged lymph nodes in children will resolve without treatment or biopsy.
        i. If lymphadenopathy persists for 4–6 weeks, surgical evaluation and possible biopsy are indicated. One exception is cat scratch disease, which leads to very large nodes that can persist for months. With a history of a cat scratch and a positive Bartonella titer, in most cases biopsy is not needed. Cat scratch disease does not always require antibiotic treatment as it is usually self-limited, although it is sometimes treated with macrolide antibiotics (e.g., azithromycin) to speed recovery.
    b. Incision and Drainage (I&D) of suppurative nodes: Antibiotics may prevent the need for drainage, but in many cases, abscess formation requires I&D. The node may be aspirated and cultured, but most often it is incised, drained, and cultured.
    c. Excision of nodes with non-tuberculosis mycobacterial infections: Surgical excision is more effective than antibiotics for the treatment of these infections. They most commonly involve submandibular and preauricular nodes. Care must be taken to avoid injury to facial nerve branches, especially the marginal mandibular branch.
    d. Surgical biopsy: May be completed with or without labs or imaging. This is diagnostic and is done as an outpatient procedure.
        i. If many nodes are enlarged, the largest or most abnormal is typically chosen for biopsy.
        ii. Any enlargement of supraclavicular nodes is generally an indication for biopsy. CXR and possibly chest CT should be done preoperatively to rule out an anterior mediastinal mass compressing the airway.
        iii. The surgical technique includes a skin incision over the node in question with dissection down to the node. Excisional biopsy is preferred, but for nodes > 2–3 cm, incisional biopsy may be safer. A minimum of 1 cubic centimeter of tissue is generally requested by pathology for evaluation. Care is taken to avoid injury to nerves and vessels, especially in the neck; specifically, the marginal mandibular branch of the facial nerve as above, and cranial nerve XI (spinal accessory) during posterior cervical triangle node biopsies.

    iv. Biopsied nodes are generally sent fresh to Pathology. Many centers have a "Lymph Node Protocol" which includes histology, infectious work-up/cultures, flow cytometry, and other testing as indicated.

  e. Fine Needle Aspiration (FNA): This currently is used more often in adult patients than in pediatrics, but in select cases may be useful to help distinguish benign from malignant disease.
  f. Core Needle Biopsy (CNB) with ultrasound guidance: This has been shown to be superior to FNA in many cases. It may be especially useful in suspected lymphoma cases with lymphadenopathy and associated mediastinal mass in which general anesthesia is contraindicated. CNB could then be done with local anesthesia and light sedation.
  g. Sentinel Node Biopsy: This technique is most commonly used in pediatrics for the staging of melanoma, but also has been described for some sarcomas and for pediatric breast cancer.

6. Outcomes:

  a. Pathology results are typically back within a few days for histology, although some cultures and other results can take weeks.
  b. Usually, no further treatment is needed if the node is reactive on pathology. Routine surgical follow-up is indicated for the biopsy site, and any further follow-up is typically with the primary care physician.
  c. Suspected or confirmed malignancies require referral to a Pediatric Hematologist-Oncologist. Survival rates for limited-stage Hodgkin and Burkitt lymphomas exceed 90%, while higher stage and other non-Hodgkin lymphomas are generally less favorable.
  d. Some infections should be referred to a Pediatric Infectious Disease specialist. These include, but are not limited to, TB, other mycobacterial infections, Actinomyces, certain fungal infections, toxoplasmosis, HIV, and resistant organisms.

## Additional Notes

In summary, lymphadenopathy is a common condition seen by pediatric surgeons. Patients are often referred by pediatricians, family physicians, and pediatric oncologists for further evaluation and potential biopsy. The surgeon's role is often one of reassurance as this is mostly a benign self-limited condition. The surgeon also plays an important role in determining which patients and lymph nodes require further intervention, such as incision and drainage of a suppurative node, or biopsy to rule out malignancy.

## Study Questions

1. Which of the following enlarged lymph nodes is the most concerning for malignancy and should essentially always be biopsied?
    a. Axillary
    b. Inguinal
    c. Posterior cervical
    d. Supraclavicular

Answer: D: Supraclavicular. Any enlargement of a supraclavicular lymph node is abnormal, and biopsy is indicated to rule out malignancy, especially in older children and teenagers. Prior to biopsy, an anterior mediastinal mass compressing the trachea must be ruled out by history and imaging.

2. What is the most effective treatment for lymph nodes with severe non-tuberculosis mycobacterial infections?
    a. Surgical excision
    b. Incision and drainage
    c. Antibiotics
    d. Expectant observation

Answer: A: Surgical excision. Antibiotics are typically not very effective treatment of significant non-TB mycobacterial lymph node infections. Complete surgical excision of involved nodes is the most effective therapy. The most commonly involved nodes are submandibular and preauricular, and therefore careful dissection is required to identify and preserve facial nerve branches, particularly the marginal mandibular branch.

3. What structure is at particular risk of injury in performing biopsy of a lymph node in the posterior cervical triangle?
    a. Internal jugular vein
    b. Carotid artery
    c. Spinal accessory nerve (Cranial nerve XI)
    d. Thoracic duct

Answer: C: Spinal accessory nerve (Cranial nerve XI). The spinal accessory nerve (Cranial nerve XI) is a purely motor nerve that innervates the sternocleidomastoid muscle and then traverses the posterior cervical triangle to innervate the trapezius. Injury to the nerve in the posterior triangle leads to winging of the scapula, which can be quite debilitating.

## Further Reading

Grant CN, Aldrink J, Lautz TB, et al. Lymphadenopathy in children: a streamlined approach for the surgeon – a report from the APSA cancer committee. J Pediatr Surg. 2021;56:274–81.

# Appendicitis

**Tara J. Loux**

**Key Points**

- Appendicitis is one of the most common surgical illnesses of childhood. Most individual cases are easily and quickly managed, especially in countries with advanced health care. However, there is substantial combined morbidity and lost productivity on a global scale.
- Trials of non-operative management demonstrate up to 10% early failure rate and up to 60% recurrent appendicitis rate in the first 2 years after therapy. While not currently standard of care, antibiotic treatment alone can be considered in certain unique situations.
- Appendectomy can be safely delayed for 12–24 h after diagnosis of appendicitis if the patient is started immediately on broad-spectrum intravenous antibiotics. The patient should be kept *nil per os* (NPO) and given resuscitative and maintenance intravenous fluids prior to operation, with electrolyte correction as needed.
- Appendectomy is curative in cases of simple appendicitis. Complicated appendicitis presumes the spread of infection into the peritoneal cavity, which requires postoperative antibiotic therapy.

1. Epidemiology

   a. Incidence: Lifetime incidence is 7% in USA. There are 60,000 pediatric cases per year in the USA. Despite improvements in diagnosis, the rate of complicated (perforated) appendicitis in children remains between 25 and 33% of

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cases and has not decreased in recent years. Appendicitis is most prevalent between 5 and 25 years of age. There is seasonal variation in USA, with presentation slightly more common in summer months. Perforation is more common in the very young (<5 years), very old (>65 years) and patients with neurologic disability.

   b. Mortality: Death from appendicitis is rare in pediatric patients, especially when otherwise healthy. There were 158 deaths reported in the US between 2010–2019 in the 18 and under age group. Twenty-five percent of these were in infants under one year of age, and the mortality rate for appendicitis in an infant may be as high as 25% due to its uncommon presentation, delay in diagnosis, immature immune system and concurrent pre-existing conditions such as prematurity.
   c. Risk factors: Male sex (M:F ratio 1.4:1), family history of appendicitis, Caucasian, or Hispanic heritage, higher median household income, urban setting, chronic constipation, cystic fibrosis.

2. Pathophysiology:

   a. Anatomy: The vermiform appendix is a small, blind-ending tubular structure that emanates from the base of the cecum. It can be intraperitoneal or retroperitoneal/retrocecal. The blood supply is from the appendiceal artery, which emanates from the ileocolic pedicle, the terminal branch of the superior mesenteric artery (Fig. 1).
   b. Etiology: Appendicitis is caused by blockage of the appendiceal orifice, whether by fecalith, enlargement of lymph tissue, tumor, or other foreign body. Build-up of pressure impedes adequate blood flow, which leads to inflammation and eventually gangrene and perforation. Perforation is thought to occur 24–48 h after the initiation of symptoms, in the absence of any intervention.
   c. Classification:

      i. Early acute appendicitis—any non-perforated appendix with signs of acute inflammation.
      ii. Perforated appendicitis—visible hole in appendix or fecalith in abdomen. Perforation can be further sub-divided into four grades:

          1. Localized or contained perforation
          2. Localized, well-circumscribed abscess
          3. Generalized peritonitis without discrete abscess
          4. Generalized peritonitis with multiple abscesses

      iii. Other appendiceal pathology—includes eosinophilic appendicitis, subacute and chronic appendicitis, lymphoid hyperplasia, appendix carcinoid or other malignancy, pinworm (Enterobius vermicularis) infection, foreign body

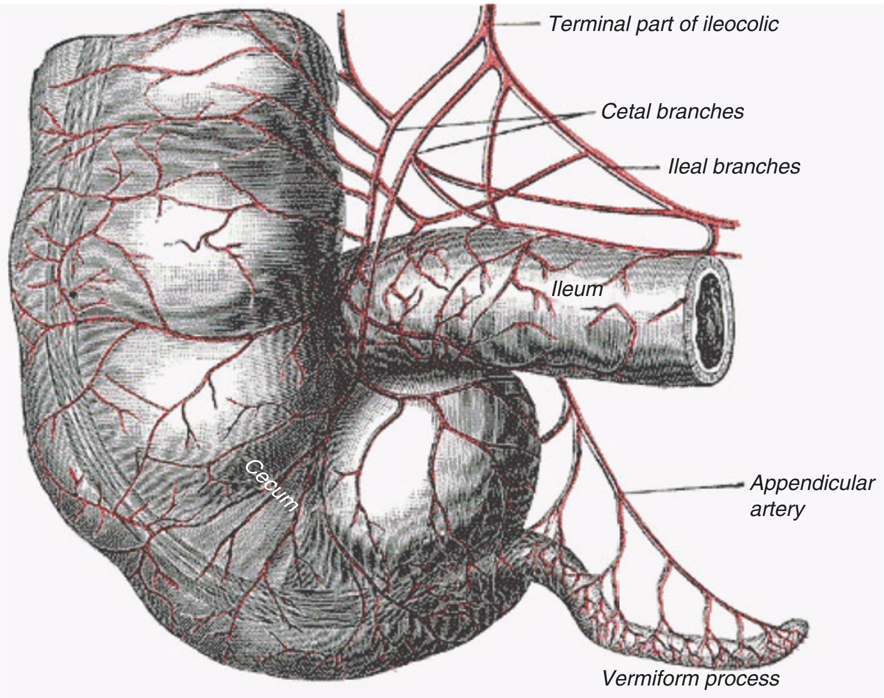

**Fig. 1** Anatomy of vermiform appendix. Source: Henry Gray (1918) *Anatomy of the Human Body*. Bartleby.com: Gray's Anatomy, Plate 536

3. Clinical Features and Diagnosis

    a. Symptoms: Classically, vague abdominal pain which is persistent, worsening, and localizes to the right iliac fossa over a short duration (12–24 h) and can be accompanied by nausea, vomiting, anorexia, and low-grade fever (<100.5 F). Early onset of high fever, diarrhea, or sporadic pain is more consistent with gastroenteritis or other diagnoses. Many younger patients (<5 years old) will present with intractable vomiting, lethargy, lack of appetite and fever, as they are less likely to be cognitively able to report or describe abdominal discomfort.
    b. Examination: A complete physical examination should be performed to rule out other potential causes of abdominal pain and document levels of activity, hydration and nutrition, abdominal or other surgical scars. In particular, note should be taken of the child's spontaneous movements, temperature, pulse, and overall appearance. A full lymphatic basin exam is imperative. An external genital examination should be done in all patients to document Tanner's stage of development and rule out inguinal hernia or testicular pathology.

Abdominal examination can be difficult in younger children. Involuntary guarding is invariably present in the right lower quadrant and should be carefully ascertained. Certain signs may be present in appendicitis to aid with diagnosis:

   i. McBurney sign—tenderness to palpation at McBurney point (a point 2/3 of the distance between the umbilicus and right anterior superior iliac spine)
   ii. Perman sign—pain felt in right iliac fossa with palpation of the left iliac fossa (also known as Perman–Rovsing sign)
   iii. Blumberg sign—rebound tenderness when releasing deep palpation in the right iliac fossa
   iv. Dunphy sign—pain felt in right iliac fossa upon coughing
   v. Psoas sign—pain produced by passive extension or active flexion of the right hip
   vi. Obturator sign—pain produced by internal rotation of the hip

c. Laboratory Tests: A complete blood count with differential, electrolyte panel, liver function panel, lipase level and urinalysis should be sent to work up the child with abdominal pain. White blood cell count will invariably be elevated, often with leftward shift, unless the patient is neutropenic for some other reason (cytotoxic chemotherapy). C-reactive protein and procalcitonin are also usually elevated. Urinalysis should be performed to rule out urinary tract causes of abdominal pain and to perform urine pregnancy screening in postmenstrual (Postmenarchal) females. Irritation of the bladder by an inflamed appendix can cause sterile pyuria (usually <20 white blood cells/high power field), without the presence of bacteria, blood, or nitrates. Typical electrolyte abnormalities (hyponatremia, hypochloremia, metabolic acidosis or hypokalemic metabolic alkalosis) can be noted in cases where there has been prolonged vomiting or low oral fluid intake.
d. Grading Systems: Several grading systems of variable sensitivity and specificity are available for the diagnosis of appendicitis in the pediatric patient with abdominal pain (for example, the Pediatric Appendicitis Score in Fig. 2, or [1]). They use a combination of variables such as character and location of pain, associated symptoms, physical examination and laboratory values to calculate a numeric score, which classifies patients into Low, Intermediate, and High suspicion groups.

   i. Low suspicion: Can safely be discharged without further workup.
   ii. Intermediate suspicion: May undergo some sort of diagnostic imaging.
   iii. High suspicion: Can be offered appendectomy, with or without confirmatory diagnostic imaging.

e. Radiologic Studies:

   i. Ultrasound should be the first radiologic test ordered. It has a high specificity but a low sensitivity and can be operator and patient dependent. The

| Pediatric Appendicitis Score | |
|---|---|
| For the presence of the following sign or symptom: | Assign the following points: |
| Pain in right lower quadrant on cough, hop or percussion | 2 |
| Tenderness to palpation of the right iliac fossa | 2 |
| Anorexia | 1 |
| Fever | 1 |
| Nausea/vomiting | 1 |
| Leukocytosis | 1 |
| Neutrophilia | 1 |
| Migration of pain to right iliac fossa | 1 |
| Total possible points | 10 |
| If PAS<5, appendicitis is highly unlikely.<br>If PAS>7, appendicitis is highly likely.<br>If PAS 5-7, likelihood is intermediate and imaging or observation may be warranted. | |

**Fig. 2** Pediatric Appendicitis Score. Source: Samuel M, "Pediatric Appendicitis Score." J Pediatr Surg. 2002; 37: 877–881, data from Elsevier complete source

appendix may not be visualized. An appendix over 6–7 mm in diameter is suspicious for appendicitis, especially if there is loss of compressibility, evidence of fecalith (shadowing stone), hyperemia of the appendiceal wall on Duplex and surrounding free fluid. Pelvic ultrasound should be ordered concomitantly in females to rule out ovarian pathology.

ii. Some centers use rapid magnetic resonance imaging (MRI) without contrast, which has a sensitivity and specificity similar to that of computed tomography (CT) with intravenous contrast, but is more costly and may not be available at scale in every institution [2].

iii. CT is quick, less expensive than MRI, has high sensitivity and specificity and is not operator dependent, but does involve the use of ionizing radiation and a quantifiable increase in the risk of hematologic malignancy. IV contrast should be administered, however, oral contrast is no longer given for CT when appendicitis is suspected due to poor tolerance, further delay in diagnosis and the potential need for subsequent urgent operation.

iv. Admission with serial abdominal examinations ("observation") can be an alternative to imaging, or appropriate for intermediate suspicion patients with a non-visualized appendix on ultrasound. Patients who undergo observation should be kept NPO and given appropriate intravenous analgesia, but should not be given antibiotics until the diagnosis has been confirmed.

   f. Differential Diagnosis: Infectious enteritis or colitis, mesenteric adenitis, intussusception, omental infarction, Crohn's disease, typhlitis, pancreatitis, abdominal trauma (accidental or otherwise), adhesive bowel obstruction, Meckel diverticulitis, ovarian or fallopian tube pathology (ectopic pregnancy, hemorrhagic ovarian follicle or ovarian torsion), urinary tract infection or nephrolithiasis, testicular pathology (torsion of testicle or epididymal appendage, infectious or inflammatory orchiditis).
   g. Occasionally, despite imaging, the diagnosis is equivocal due to a variety of factors, and diagnostic laparoscopy may be offered to clarify the diagnosis. If a normal appendix is found, it should be removed to eliminate this diagnosis in the future. However, if evidence of Crohn's disease is found on laparoscopy, the appendix should NOT be removed, as there is a high rate of appendiceal stump leak and enterocutaneous fistula. Standard therapy for Crohn's disease should be initiated. Contraindications to laparoscopy include acute abdomen with hemodynamic abnormality or history of multiple previous intraabdominal operations.
   h. It is accepted that a normal appendix may be found at operation, despite a thorough preoperative workup. Prior to the availability of cross-sectional imaging, the acceptable number was as high as 25%. Currently, preoperative diagnostic sensitivity and specificity are around 96%, such that a normal appendix finding should occur in less than 4% of cases.

4. Treatment

   a. Initial Measures:

      i. Patient is made NPO and given 20 mL/kg 0.9% saline bolus, followed by appropriate maintenance intravenous fluids.
      ii. A broad-spectrum antibiotic is started. Cefoxitin 40 mg/kg up to 2 g is appropriate unless complicated appendicitis is highly suspected or confirmed on preoperative imaging.
      iii. Intravenous analgesia with morphine 0.05 mg/kg and/or acetaminophen 15 mg/kg is given for patient comfort. Analgesia should not be withheld pending surgical consultation.
      iv. Patients with severe sepsis or septic shock should be stabilized medically prior to operation.

   b. Subsequent Measures:

      i. Operative therapy is the standard of care and should be accomplished in most cases where the patient can safely undergo general anesthesia.
      ii. Operation can safely be delayed 12–24 h in patients who are receiving appropriate antibiotic therapy without risk of perforation while in hospital.
      iii. In cases where the patient is not a candidate for immediate general anesthesia, intravenous antibiotic therapy should be continued until oral intake is re-established, and oral antibiotic therapy continued until the patient experiences complete resolution of symptoms. Occasionally,

symptoms continue despite antibiotic therapy and antibiotics must be continued until operation is planned.

iv. If a well-formed abscess is present, percutaneous drainage placement should be offered. Interval appendectomy may be offered 6–8 weeks thereafter.

c. Preoperative Preparation: Have patient empty bladder immediately prior to operation or perform Credé maneuver after induction of anesthesia. General anesthesia is necessary for laparoscopy, however, spinal anesthesia can be sufficient for right lower quadrant incision. Foley catheter should not be necessary in most cases. Clip abdominal hair in older patients. Tuck left arm (or both arms). Ensure antibiotic is dosed or re-dosed 15 min to 1 h prior to incision as per standard guidelines. Prep the entire abdomen with chlorhexidine or betadine solution.

d. Operative Treatment: Appendectomy can be performed open through a right lower quadrant (Rocky–Davis) or lower midline incision. However, studies have shown significantly improved short and long-term postoperative outcomes with laparoscopy, including decreased postoperative pain, quicker return to activity, fewer postoperative surgical site infections and fewer adhesive bowel obstructions. Laparoscopy can be conducted via the traditional three-port method, with SILS (single-incision multichannel laparoscopy), or with SISI (single-incision single-instrument) laparoscopy.

Regardless of how it is performed, appendectomy consists of the following steps:

i. Careful separation of omentum, adjacent bowel loops and (if applicable) adnexal structures from the inflamed appendix. This may also entail mobilization of the cecum and right colon to gain access to the appendix.
ii. Creation of a window in the mesoappendix with ligation of the appendiceal arterial pedicle.
iii. Ligation of the base of the appendix flush at its juncture with the cecum, taking care not to narrow the entrance of the terminal ileum into the cecum, nor leave behind any portion of the base that might cause stump appendicitis.
iv. Removal of all fluid, infected debris, fecalith, and purulence from the abdominal cavity.

e. Postoperative Treatment:

Patients with *uncomplicated appendicitis* can safely be discharged from the recovery room after laparoscopic surgery. No further antibiotic therapy is necessary when no perforation is found operatively. They should be allowed diet as tolerated. Pain control with oral acetaminophen and ibuprofen is sufficient. Activity, including normal bathing, should be quickly liberalized. Most patients can return to school or light duty within 3–5 days and regular activity in 2 weeks.

*Perforated appendicitis* presumes spread of infection within the abdominal cavity and requires postoperative antibiotic therapy. Most surgeons agree that patients with perforative appendicitis should receive at least 5 days of antibiotic therapy. In this author's experience, antibiotics should be continued until symptoms are fully resolved, including abdominal pain, diarrhea, and fever. There is clear evidence that patients can be safely switched to oral therapy and discharged home once oral intake is tolerated (a full meal). Average hospital stay is 4–5 days. A Dutch trial comparing two days of total antibiotic therapy versus five days in patients 8 years old or greater was scheduled to finish recruiting in 2020 and will hopefully result in the near future.

Most patients with perforated appendicitis will have diarrhea initially and this does not mean they have resumed normal bowel function. A nasogastric tube should be placed to low continuous suction if they have more than one episode of vomiting postoperatively, or if they experience bilious emesis. Oral intake is the best predictor of an improving child. If oral intake has not reached or is not expected to reach normal levels by postoperative day 7, total parenteral nutrition (TPN) should be started via peripherally inserted central venous catheter (PICC). Additionally, at this time postoperative imaging should be obtained, to start with ultrasound looking for fluid collection. Waiting until day 7 to start TPN and perform imaging has shown to significantly reduce the amount of unnecessary TPN and postoperative imaging and does not increase the risk of other complications.

f. Complications: Complication rate after appendectomy for uncomplicated appendicitis should be 3% or less. Complication rate after appendectomy for ruptured appendicitis can range from 10 to 25%. Complications include bleeding, intraabdominal abscess, wound infection or dehiscence, ileus, bowel obstruction, appendiceal stump leak, enterocutaneous fistula, antibiotic-associated diarrhea or *Clostridium difficile* colitis and infertility in female patients from scarring of the fallopian tubes.

## Additional Notes

Significant attention in recent years has been given to management of appendicitis with antibiotics alone, especially under the influence of the 2020 SARS-coV2 pandemic limitations on hospital, operating room and intensive care capacities. The few studies published with pediatric data are retrospective or non-randomized, and have reported an average 10% early failure rate of non-operative therapy, and 25% rate of recurrent appendicitis in the first year. Some studies show decreased number of disease-related activity limitation days in non-operative therapy groups in the short term. No study has reported follow-up in children for more than 2 years at this point. Given current antibiotic stewardship priorities, the ease and relative safety of performing appendectomy in a healthy child versus an older adult, and the much longer ensuing life span of a child, this author's opinion is that non-operative management of appendicitis in children is inferior treatment and should only be offered in select circumstances, followed by the option of outpatient interval appendectomy. Clinical trials to study this in more detail are currently ongoing [3].

## Study Questions:

1) Question 1: You remove a non-perforated appendix from an otherwise healthy 8-year-old child with a one-day history of symptoms. The patient is discharged home the same day, and the pathology report resulting 5 days later notes the presence of *Enterobium vermicularis*. What should be done about this finding?

   a. No further intervention is required.
   b. Prescribe 7 days of oral amoxicillin/clavulanate.
   c. Admit to the hospital for 7 days of intravenous piperacillin/tazobactam.
   d. Prescribe oral mebendazole 100 mg, two doses separated by two weeks.

Answer to Question 1: d) Pinworm is found incident with appendicitis in up to 4% of specimens. Treatment should include members of the entire household of the patient.

2) What is the appropriate antibiotic treatment regimen in a patient with complicated appendicitis?

   a. Ceftriaxone 50 mg/kg up to 2 g and metronidazole 30 mg/kg up to 1 g intravenously every 24 h for 5 days postoperatively.
   b. Treatment regimen is dependent on patient characteristics, signs, and symptoms.
   c. Piperacillin/tazobactam 100 mg/kg up to 3.375 g, intravenously every 6 h for 7 days.
   d. Sulfamethoxazole/trimethoprim 4 mg/kg up to 160 mg trimethoprim orally twice daily for 7 days.
   e. Gentamicin 2.5 mg/kg (following peak and trough levels) and clindamycin 13 mg/kg (up to 900 mg) intravenously every 8 h for 6 days postoperatively.

Answer to Question 2: b) Patients with complicated appendicitis remain hospitalized on intravenous antibiotics until they are able to tolerate oral intake (a full meal). Options a, c, d, and e are all potential treatment regimens. The most cost-effective intravenous treatment regimen is ceftriaxone and metronidazole, given once daily. Alternatives include piperacillin/tazobactam, clindamycin + gentamicin, ciprofloxacin + metronidazole, or a carbapenem, depending on the individual patients' allergy profile. Once oral intake has stabilized, the patient can be switched to oral antibiotics and discharged home. Appropriate oral antibiotics include amoxicillin/clavulanate 22 mg/kg up to 875 mg amoxicillin every 12 h, sulfamethoxazole/trimethoprim, or ciprofloxacin 10 mg/kg up to 500 mg every 12 h + metronidazole 10 mg/kg up to 500 mg every 8 h. Most treatment protocols agree that therapy, whether oral or intravenous, should continue for a minimum of five days postoperatively, AND until full resolution of abdominal pain, diarrhea, and fever.

3) Question 3: You are called to see a 5-year-old patient with acute lymphoblastic leukemia in the oncology ward who has developed right lower quadrant pain and fever. He is currently undergoing induction therapy. The child's absolute neutrophil count (ANC) is 40 cells/uL. The pain began in the periumbilical region

before radiating to the right lower quadrant. The child has positive McBurney and Perman signs. There is no evidence of diffuse peritonitis. An ultrasound shows a blind-ending, tubular, non-compressible structure in the right lower quadrant measuring 8 mm in diameter with increased blood flow in the walls and a small amount of free fluid. What do you recommend?

a. Emergent appendectomy.
b. CT of the abdomen and pelvis with intravenous and oral contrast.
c. Broad-spectrum intravenous antibiotics, NPO, intravenous fluids, analgesia, and nutrition with the removal of the appendix when the ANC is greater than 500 cells/uL.
d. Broad-spectrum intravenous antibiotics and percutaneous drain placement.

Answer to Question 3: c). This patient is an excellent candidate for interval management of acute appendicitis. Any operation conducted in this patient with severe neutropenia will carry an extremely high risk of wound healing complications. Cases in which it would be appropriate to consider delay of operation include concomitant presentation of influenza or respiratory syncytial virus, severe neutropenia or thrombocytopenia, severe sepsis with shock or other organ failure, diabetic ketoacidosis, or other uncontrolled medical diagnoses which might increase the risk of general anesthesia or operative complications. Interval management is also best for patients who have a well-formed abscess which is amenable to percutaneous drainage. Some surgeons recommend interval management for any patient suspected of having ruptured appendicitis; while not universally accepted, it has not definitively been proven inferior. In the case of severe neutropenia, typhlitis (inflammation of the cecum and right colon) should be suspected in a child with evidence of right lower quadrant pain, hemodynamic abnormalities and peritonitis and should be confirmed by CT scan with IV contrast. Surgical intervention for typhlitis is associated with a very high mortality rate.

## References

1. Tam D, Vazquez H. Calculated decisions: pediatric appendicitis risk calculator (pARC). Pediatr Emerg Med Pract. 2019;16(9):CD5–CD6. https://www.ebmedicine.net/media_library/files/Pediatric-Appendicitis-CD.pdf
2. Anderson KT, et al. Approaching zero: implications of a computed tomography reduction program for pediatric appendicitis evaluation. J Pediatr Surg. 2017;52:1909–15.
3. Minneci P, et al (Midwest Pediatric Surgery Consortium). Multi-institutional trial of nonoperative management and surgery for uncomplicated appendicitis in children: design and rationale. Contemp Clin Trials. 2019;83:10–7.

## *Further Reading*

Yousef Y, Youssef F, Homsy M, et al. Standardization of treatment for pediatric perforated appendicitis improves outcomes. J Pediatr Surg. 2017;52:1916–20.

# Non-malignant Splenic Disorders

**Marc S. Lessin**

**Key Points**

1) The spleen functions to remove senescent erythrocytes from the circulation and to make immune globulin and other proteins involved in the immune system.
2) Hereditary spherocytosis (HS) is an inherited hemolytic anemia caused by abnormalities in spectrin and other proteins which affect the structural integrity of the erythrocyte, which then become entrapped within the spleen leading to anemia and splenomegaly.
3) Idiopathic thrombocytopenic purpura (ITP) is marked by thrombocytopenia due to antibody-mediated destruction of platelets primarily of splenic origin
4) Thrombotic thrombocytopenic purpura (TTP) is a severe disease due to platelet aggregation in the microvasculature. TTP is marked by fever, thrombocytopenia, hemolytic anemia, neurological symptoms (e.g., headache, stroke), and renal insufficiency.
5) Wandering spleen is a condition in which the spleen may undergo torsion and requiring splenopexy.
6) Splenic cysts may be congenital, post-traumatic, parasitic, or infectious. Parasitic cysts due to Echinococcus may be treated with percutaneous infusions of normal saline or alcohol or splenectomy. Asymptomatic non-parasitic cysts <4 cm do not require treatment.

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7) Splenectomy may be indicated for splenomegaly, anemia (hereditary spherocytosis), thrombocytopenia (idiopathic thrombocytopenic purpura/thrombotic thrombocytopenic purpura), splenic sequestration (sickle cell disease), pyruvate kinase deficiency, or symptomatic splenic cysts.

The spleen is a lymphoid organ that functions as a filtering system for the hematopoietic system. The spleen is also a reservoir with normally one-third of the platelets pooled within organ. It also produces immunoglobulin in particular IgM, and is involved in maturation of B-cells required for IgG, and peptides involved in the immune system, including the opsonins tuftsin and properdin. The spleen removes senescent erythrocytes and breaks them down. In hereditary spherocytosis, the erythrocytes lack proteins, including spectrin which prevents erythrocytes from deforming to pass through the splenic vascular, resulting in their entrapment in the splenic parenchyma leading to anemia, increased reticulocytes count, splenomegaly, jaundice, and pigmented gallstones. In immune thrombocytopenic purpura (ITP) and thrombotic thrombocytopenic purpura, the spleen makes anti-platelet antibody resulting in potential bleeding secondary to thrombocytopenia.

A: Anatomy

1. The spleen is a hematologic organ that is located in the posterior left upper quadrant of the abdomen and partially protected by the lower ribs (9th–11th). Splenomegaly may result in the lower spleen extending far below the lower rib.
2. The main arterial blood supply of the spleen is from the splenic artery, which is a branch of the celiac trunk. Blood supply to the upper pole is from short gastric arteries, which are branches of the left gastroepiploic artery.
3. Venous drainage of the spleen is through the splenic vein, which runs along the posterior surface of the pancreas. The inferior mesenteric vein drains into the splenic vein, which then join the superior mesenteric vein to form the portal vein.
4. The spleen is suspended by the gastrosplenic, splenorenal, and splenophrenic, and splenocolic ligaments.

## Hereditary Spherocytosis

A. Epidemiology

Hereditary spherocytosis (HS) is a genetically inherited hemolytic anemia. Seventy-five percent occur as the result of autosomal dominant inheritance and 25% are autosomal recessive with new mutations being identified. The prevalence is approximately 1 in 5000. It occurs in all racial groups but the majority (75%) are in those of European descent. Its prevalence is equal in male and females.

1) Prevalence and inheritance:
    (i) Occurs in all racial groups but is most common in northern Europeans.
    (ii) Seventy-five percent of cases are of dominant inheritance and 25% are recessive with new mutations common.
    (iii) Many mutations may cause phenotypic HS, with mutations often unique to each family.

B. Pathophysiology

1) Peripheral blood smear will show spherocytes. The spherocytes have mutations of the spectrin protein and other proteins involved in the structural integrity and function of spectrin. The deficiency leads to a remodeling of the usual biconcave disk into cells that resemble spheres.
2) These protein defects lead to a weakened cell membrane of the erythrocyte which results in decrease in deformability. The red blood cells become trapped as they pass through the splenic pulp, which then leads to their breakdown.
3) The spherocytes have a shortened lifespan and their destruction leads to anemia.
4) As more membrane is lost, the limit of the RBC membrane stretching is reached, and lysis occurs.

C. Classification Schemes:

HS can be classified as mild, moderate, or severe based on baseline hemoglobin, degree of reticulocytosis, and resting bilirubin levels. These clinical classifications correlate with degree of osmotic fragility (severe has more fragile RBCs) and spectrin content (mild having near-normal spectrin, severe with less than 50% of normal value).

D. Clinical Presentation

1) Neonates with HS often have prolonged jaundice. Anemia during the first year of life may necessitate transfusions.
2) Patients with mild to severe forms with varying degrees of anemia, jaundice, increased reticulocyte count, variable splenomegaly, and often evidence of gallstones (pigmented from RBC breakdown).
    i. In mild cases of HS there is a compensated hemolytic anemia with normal or near-normal hemoglobin and mildly increased reticulocyte counts. These patients are usually asymptomatic and often only come to light during an illness (such as infectious mononucleosis), pregnancy or physical stress.
    ii. Moderately affected patients are generally asymptomatic but may become symptomatic when stressed.
    iii. Severely affected patients have significant anemia and are transfusion dependent. Peripheral smears in these patients show spherocytes as well as bizarrely shaped cells (poikilocytes). Growth failure, and delayed

sexual maturation are common. These patients are at risk for aplastic crises.

E. Diagnosis

1) Physical examination may demonstrate splenomegaly. Massive splenomegaly may be present with the spleen palpable to the left anterior superior iliac spine.
2) The CBC with differential shows spherocytes and usually there is a family history. A positive osmotic fragility test confirms the diagnosis.
3) Osmotic fragility testing is performed by exposing control and suspected HS cells to increasingly hypotonic sodium chloride concentrations. Normal cells are able to swell and increase their volume since they are starting as a normal biconcave disc. HS cells already have a decreased surface to volume ratio (spheres) and thus reach their limit at a higher concentration of sodium chloride compared to normal cells.
4) Additional laboratory evaluation reveals reticulocytosis, indirect hyperbilirubinemia, and variable anemia. Baseline hemoglobin levels, bilirubin levels and reticulocyte counts are helpful to document a significant change in the event of illness.
5) Differential diagnosis includes autoimmune hemolytic anemia, Heinz body anemias, or patients presenting in aplastic crises.
6) Ultrasound is the test of choice for evaluating the size and volume of the spleen.

F. Treatment

1) The majority of affected individuals do not require splenectomy.
2) If splenomegaly is present, contact sports during periods of significant enlargement should be avoided.
3) Patient should be counseled concerning indications, risks, and benefits of splenectomy.
4) Signs and symptoms of gallstones and gallbladder disease should be discussed as well as indications for cholecystectomy.
5) Splenectomy for HS is indicated for severe anemia, need for recurrent transfusion, painful splenomegaly, growth retardation, and desire to participate in contact sports in those with splenomegaly.
    i. Splenectomy ameliorates the anemia, hyperbilirubinemia, and reticulocytosis of most patients with HS. It does not eliminate spherocytes in the peripheral smear. The risk of post splenectomy sepsis must be considered in any patient undergoing splenectomy. Splenectomy for HS should not be done before 5-years of age.
    ii. Post splenectomy sepsis (OPSI) is a serious and potentially fatal complication after removal of the spleen. Splenectomized individuals are particularly at risk for severe bacterial infections usually due to Gram-positive

encapsulated organisms such as *Streptococcus pneumoniae*, *Neisseria meningitidis,* and Haemophilus influenza type b.

iii. All patients undergoing elective splenectomy should be vaccinated at least 2 weeks before splenectomy. Most children receive streptococcal and haemophilus vaccinations routinely but not *Neisseria meningitidis*. It is unclear if the risk of overwhelming sepsis lessens over time. Children undergoing splenectomy should receive prophylactic penicillin. The duration of treatment is controversial, with some advocating treatment until age 18.

iv. The preferred approach to splenectomy is laparoscopic. Partial splenectomy is an option. Patients with gallstones should undergo concomitant cholecystectomy. Attempts should be made to identify accessory spleens, which are most commonly in descending order located with the splenic hilum, gastrosplenic ligament, gastrocolic ligament and along the splenic vein to the level of the portal vein.

## Immune Thrombocytopenia (ITP)

A. Epidemiology

Most common autoimmune disorder affecting blood elements estimated to affect 1 in 10,000 children which may be an underestimate as asymptomatic children may not seek medical attention. In younger children, the sexes are equally affected. In adolescents, the female to male ratio is two to one.

B. Etiology

IgG autoantibodies interact with membrane glycoproteins on platelets or megakaryocytes, which are then cleared by the reticuloendothelial system.

C. Pathophysiology:

  a. Idiopathic thrombocytopenic purpura (ITP) is marked by thrombocytopenia occurring due to destruction of antibody sensitized platelets by the reticuloendothelial system (RES). Process is not well understood.
  b. Classification Schemes:
    i. Type I: Newly diagnosed within 3 months.
    ii. Type II: Persistent of 3–6 months duration.
    iii. Type III: Chronic—greater than 6 months duration.

D. Clinical Features: Classical presentation is that of a healthy child who presents with bruising, petechiae, epistaxis, or bleeding from the gums, and thrombocytopenia. Intracerebral hemorrhage is a rare complication requiring emergent splenectomy.

E. Diagnosis

  a. Physical Examination: Examination for bruising of the lower extremities. Splenomegaly is generally absent.

  b. Laboratory Tests:
     i. Complete blood count with differential demonstrating thrombocytopenia with normal white blood cell count and hemoglobin level, blood type and Coombs test, quantitative immunoglobulins, anti-nuclear antibody, HIV, viral testing including hepatitis, Epstein Barr virus, and cytomegalovirus in appropriate circumstances.
     ii. Bone marrow biopsy showing normal or increased megakaryocytes.
     iii. Platelet clumping caused by EDTA or cold agglutinins or megaplatelets which are miscounted by automated machines can give falsely low platelet counts.
  c. Differential diagnosis: ITP is a diagnosis of exclusion. No commercially available test is available for anti-platelet antibodies. Etiologies that should be excluded are medication-induced thrombocytopenia, viral infections such as human immunodeficiency virus, Epstein Barr virus (mononucleosis), cytomegalovirus, and rubella. *Helicobacter pylori* has been associated with thrombocytopenia in rare instances.
  d. Radiographic Studies: No radiographic studies are indicated. Ultrasound should demonstrate a normal-sized spleen (absence of splenomegaly).

F. Treatment: Depends on severity of illness.
  a. Initial Measures: Asymptomatic patients with platelet counts higher than 50,000 can be observed. Those with platelet counts between 30,000 and 60,000 that are asymptomatic may be observed. Symptomatic patients with platelet count below 50,000 and those with platelet counts below 30,000, even if asymptomatic require treatment.
  b. Subsequent Measures: Initial treatment is high dose prednisone (1 mg/kg/day). Platelet counts will typically increase in about two-thirds within 3 weeks of treatment.
  c. Those that fail to increase platelets with prednisone or remain symptomatic should receive intravenous immune globulin.
  d. Platelet transfusion is only indicated for significant bleeding as the destruction of platelets is rapid due to circulating antibody. Those that fail medical therapy or experience cerebral hemorrhage should undergo splenectomy. Cerebral hemorrhage is an indication for platelet transfusion and emergent splenectomy.
  e. Preoperative Preparation: Splenectomy should be reserved for patients with significant risk of bleeding where medical therapy has either failed or the patient has developed unacceptable toxicity, and should be delayed until after age 5-years-old due to risk of post-splenectomy sepsis. Patients should be vaccinated against Haemophilus Influenza Type b, Pneumococcus, and Neisseria Meningitidis. Vaccinations ideally should be given 2–3 months before splenectomy and no less than 2 weeks prior. Intravenous immune globulin should be given within 3 days of splenectomy. This has been shown

to transiently increase platelet counts and increase the efficacy of platelet transfusion.

f. Operative Treatment: The preferred treatment is laparoscopic splenectomy. Platelet transfusions are generally unnecessary. Check for accessory spleens.
g. Postoperative Treatment: Antibiotic treatment after splenectomy is controversial. Current recommendations are that amoxicillin or penicillin be administered daily until age 16 or at least until age 5 or 3 years after splenectomy. Clear instructions need to be given, including antibiotic supply and instructions to immediately start antibiotics with the onset of any febrile illness or illness associated with rigors, chills, sore throat, malaise, myalgias, diarrhea, or vomiting.
h. Complications to Watch for: Failure rate of laparoscopic splenectomy is approximately 25% at 5 years.

    i. Check peripheral smear for Howell-Jolly bodies—present in the asplenic patient. Presence of Howell-Jolly bodies implies splenic tissue remains.
    ii. If Howell-Jolly bodies present, evaluate for missed accessory spleen. This is generally done with nuclear medicine liver-spleen scan. CT may also be useful to identify location and anatomy.
    iii. Overwhelming post-splenectomy infection (OPSI) is a life-threatening complication with most infections occurring within 2 years but over 40% occurred after 5 years. Mortality associated with OPSI is almost 50% with significant potential morbidities in survivors including loss of digits and limbs.

## Thrombotic Thrombocytopenic Purpura (TTP)

A. Epidemiology: TTP is a rare disorder marked by waxing and waning episodes of organ and tissue ischemia secondary to reversible platelet aggregation in the microvasculature. TTP is usually acquired, although inherited chronically relapsing forms also occur.

    1) Incidence: One to four per million with a slight female predominance, and peaks between ages 30–40-years. Fifteen percent of patients have an underlying condition such as infection, pregnancy, collagen vascular disorders, and drugs have been implicated, including cyclosporine, tacrolimus, quinine, and clopidogrel. No geographic or racial disposition has been identified. More common in pregnancy and patients with human immunodeficiency virus (HIV).

B. Pathophysiology: Characterized by thrombi containing aggregated platelets with little inflammatory response. Lesions usually involve small vessels (arterioles and capillaries). Brain, abdominal organs, and the heart are most commonly involved. Microthrombi contain large amounts of von Willebrand factor (VWF). An abnormal von Willebrand protein is thought to be directly involved in the pathology. Deficiency or loss of function of protein ADAMTS13 has been implicated. Acquired TTP is often secondary to an autoantibody against ADAMTS13 which disrupts its function.

C. Clinical features: Presentation to be considered is the pentad of thrombocytopenia, microangiopathic hemolytic anemia, neurologic symptoms, fever, and renal abnormalities.

D. Diagnosis—clinically based. Evaluation should not delay treatment as untreated TTP associated with 90% mortality.

   1) Symptoms: Usually acute with bleeding and rapidly progressive neurologic symptoms including headache, coma, stupor, seizures, and hemiparesis. Fever, anemia, bleeding, renal abnormalities, jaundice, fatigue, nausea and vomiting, abdominal pain, chest pain, cardiac arrhythmias, and joint or muscle pain are variably present. Without treatment the mortality rate may reach 90%.
   2) Examination: Bleeding, ecchymosis from impaired platelet function, neurological symptoms, and edema from renal insufficiency.
   3) Laboratory Tests: Rule out disseminated intravascular coagulation including prothrombin time (PT), partial thromboplastin time (PTT), INR, and fibrin degradation products. Complete blood count, chemistries to include lactate dehydrogenase, haptoglobin, and reticulocyte count. Peripheral smear will demonstrate schistocytes, nucleated red blood cells, increased LDH, decreased haptoglobin, and increased reticulocyte count. Diagnosis confirmed by measurement of ADAMTS13 activity or antibodies against it.

E. Treatment

   1) Initial Measures: The mainstay of treatment is plasma exchange or in cases of congenital TTP, fresh frozen plasma is transfused to replace the deficient enzyme. Early diagnosis and treatment have resulted in reduced mortality from what was 85–100% to 10–30%. Plasma exchange is continued until platelet count normalizes.
   2) Subsequent Measures: Relapse rates are noted in about one-third within 30 days of the initial response. Chemotherapy agents such as rituximab, vincristine, cyclophosphamide, and splenectomy are treatments for relapse.

## Pyruvate Kinase Deficiency/Glucose-6-Phosphate Dehydrogenase Deficiency

Pyruvate kinase deficiency (PKD) and glucose-6-phosphate dehydrogenase deficiency (G6PD) are hereditary conditions associated with hemolytic anemias.

A. Epidemiology: PKD is the most common enzymatic deficiency resulting in hemolytic anemia due to shortened lifespan of the red blood cell.

a. Incidence: PKD is found primarily in the Caucasian population with an incidence of 51 per million. A particularly high proportion is noted in the Pennsylvania Amish population. G6PD is postulated to affect more the 400 million worldwide.
b. Etiology: Both G6PD and PKD hemolytic anemias are due to an enzyme deficiency in the erythrocyte. PKD is autosomal recessive condition. G6PD is an X-linked condition typically seen in those of African or Mediterranean descent and is therefore more common in males.

B. Pathophysiology: Pyruvate kinase is involved in the production of adenosine triphosphate (ATP). The lack of ATP results in defects in the membrane integrity of the erythrocyte, which are destroyed by the spleen by phagocytosis. G6PD deficiency affects the pathway in the production of NADPH in oxidative phosphorylation. NADPH prevents oxidative stress in the erythrocyte. Red blood cells are subject to destruction when faced with situations of oxidative stress, including hypoxia, exposure to certain foods (fava beans), medications, and environmental factors.

C. Clinical features: PKD is associated with chronic hemolysis and need for transfusion. Both PKD and G6PD can result in hyperbilirubinemia in the newborn. G6PD is associated with and acute episodic hemolysis and is usually not chronic.

D. Diagnosis

a. Symptoms: PKD should be suspected in one with a family history suffering active hemolysis in the absence of an autoimmune disease, red cell membrane defect, or hemoglobinopathy in the setting of family history. In the newborn period, it results in hyperbilirubinemia, often severe. G6PD is usually asymptomatic.
b. Laboratory Tests: PKD can be confirmed based on gene testing and low pyruvate kinase enzyme activity. G6PD may show Heinz bodies on the peripheral smear. Haptoglobin may be saturated and in severe cases, free hemoglobin can be seen in the urine. The diagnosis of G6PD show enzyme levels $\leq 10\%$ of normal.

E. Treatment

a. Initial Measures: PKD deficiency in the newborn may result in severe hyperbilirubinemia requiring phototherapy and/or exchange transfusion. Transfusion is usually not required in children maintaining a hemoglobin level of 7–9 g/dl. Treatment of G6PD involves avoidance of exacerbating facts that lead to oxidative stress, which result in the breakdown of the red corpuscle.
b. Subsequent Measures: Splenectomy has been shown to be helpful in decreasing transfusion requirements in PKD and is recommended in patients that require frequent transfusions or that do not tolerate anemia. Splenectomy usually results in a rise in hemoglobin level of 1–3 g/dl. Splenectomy is not indicated nor is it effective in G6PD.

## Splenic Sequestration (see chapter on sickle cell disease)

Splenic sequestration occurs secondary to the trapping of red blood cells in the spleen. It may have rapid onset. Children may present in shock, which can be fatal. Young children with hemoglobin SS or hemoglobin S beta thalassemia 0 are affected prior to their spleens becoming fibrotic, while individuals with milder forms of disease present later in life. Patients present with a rapidly enlarging spleen, low hemoglobin, and often a brisk reticulocytosis. Prompt fluid resuscitation and packed red blood cells are necessary and lifesaving. Many will have a second sequestration event. Severe or recurrent episodes require splenectomy.

## Wandering Spleen

A. Epidemiology:

  1) Incidence: Rare condition seen in children and primarily young women.
  2) Etiology: Failure to form normal peritoneal attachments securing the spleen. A second theory is that hormonal changes in women lead to abdominal laxity, which elongates the splenic pedicle placing the spleen at risk for torsion.

B. Diagnosis

  1) Symptoms: Intermittent abdominal pain with splenomegaly due to venous congestion.
  2) Physical Examination: The spleen may be palpated as a mobile mass in the left upper abdomen.
  3) Radiographic Studies: CT with contrast demonstrates the spleen in an abnormal position.

C. Treatment

  1) Initial Measures: Symptomatic patients should undergo splenopexy. Torsion with splenic infarction may require splenectomy.
  2) Preoperative Preparation: Patients should be vaccinated against Gram-positive encapsulated organisms prior to any procedure which poses a risk for splenectomy.
  3) Operative Treatment: Laparoscopic splenopexy is the treatment of choice.

## Splenic Cysts/Splenic Abscess

A. Epidemiology

  1) Etiology: Splenic cysts may be post-traumatic, congenital, or parasitic. True cysts with an epithelial lining account for about 10%. Non-parasitic pseudo-

cysts account for 70–80% of cystic disease of the spleen. Splenic abscess usually occurs as the result of hematogenous spread. It may be related to infection of a post-traumatic splenic cyst.

B. Pathophysiology:

  1) Anatomy: True cysts have an epithelial lining and are considered congenital. Pseudocysts lack a true epithelial lining.
  2) Classification Schemes: Parasitic and non-parasitic. Non-parasitic cysts are further divided into true cysts (epithelial lined) and pseudocysts.

     i. Parasitic cysts are usually hydatid in nature, resulting from Echinococcus.
     ii. True cysts with an epithelial lining account for about 10%.
     iii. Pseudocysts are usually associated with a previous history of trauma. They lack an epithelial lining.

C. Clinical features: Hydatid cysts occur in children that have visited or live in endemic areas. Pseudocysts are usually related to splenic trauma.

D. Diagnosis

  1) Symptoms: Hydatid cysts occurs in areas of endemic disease. True cysts are usually asymptomatic and discovered incidentally. True cysts and pseudocysts may result in the feeling of abdominal fullness, early satiety, pleuritic chest pain, shortness of breath, back pain and/or shoulder pain. Pseudocysts are often associated with a previously history of trauma, particularly in conjunction with subcapsular hematoma. Splenic cysts rarely rupture. Occasionally, a splenic cyst may become infected, resulting in a splenic abscess. Splenic abscess manifests as left upper quadrant pain, pleuritic chest pain, peritonitis, and fever. These can occur post-traumatic but primarily occur in immunocompromised individuals.
  2) Examination: Splenic cysts may occasionally be palpable.
  3) Laboratory Tests: Hydatid cysts may be confirmed by serologic testing. Epidermoid cysts test positive for CA19-9 and carcinoembryonic antigen but do not have malignant potential.
  4) Radiographic Studies: Radiographic imaging with ultrasound or CT should be performed. Hydatid cysts will reveal calcifications. Splenic cysts appear as a smooth, unilocular lesions, occasionally with calcification.

E. Treatment

  1) Initial Measures: Splenectomy is the treatment of choice for hydatid cysts. Spillage needs to be prevented as spillage of cyst contents into the peritoneal space may result in anaphylaxis. Hydatid cysts can also be treated by injection of hypertonic saline or alcohol. Antiparasitic agents of mebendazole and albendazole are the only effective anthelmintics. Asymptomatic true cysts and pseudocysts may be observed if <4 cm. Those that are symptomatic or

>4 cm may be treated by percutaneous drainage but recurrence is high. Splenic abscess is treated by percutaneous drainage.

2) Subsequent Measures: Partial splenectomy or cystectomy with omental onlay is usually effective for non-parasitic cysts.

## Additional Notes

The spleen is an important hematological and lymphatic organ which is both a blood and platelet reservoir and a filtration system to remove red blood cells from the circulation. The spleen has important immunological function as a source of immune globulins and peptides which stimulate the immune system. It is not an essential organ but its removal puts one at risk for several and sometimes fatal infectious complications. The conditions outlined in this chapter sometimes require splenectomy. Splenectomy should not be taken lightly as overwhelming post-splenectomy infection (OPSI) is associated with severe morbidity and significant mortality. Procedures to salvage the spleen such as partial splenectomy should be considered whenever feasible.

## Study Questions

1) Splenectomy is (sometimes) indicated for all of the following conditions EXCEPT:

   a. Hereditary spherocytosis
   b. Splenic sequestration
   c. Idiopathic thrombocytopenic purpura
   d. Glucose-6-phosphate dehydrogenase deficiency
   e. Pyruvate kinase deficiency

Answer to Question 1: (d): Splenectomy may be necessary for all the conditions listed but is rarely indicated for G6PD. Cells with G6PD deficiency are susceptible to oxidative stress which is precipitated by exposure to certain foods, medications, or environmental factors. Primary treatment is avoidance of factors that lead to oxidative stress and destruction of the red blood cells. All other conditions listed may benefit from splenectomy as a second-line treatment.

2) Which of the following is most likely to require splenectomy for idiopathic thrombocytopenic purpura (ITP)?

   a. Sixteen-year-old patient recently diagnosed with ITP who refuses to take prednisone
   b. Twelve-year-old patient with intracerebral hemorrhage

c. Four year old with acute thrombocytopenia associated with a recent viral illness
d. Seventeen year old with ITP during pregnancy

Answer to Question 2: (b): Intracerebral hemorrhage in a patient with ITP is an indication for emergent splenectomy. All the other conditions listed may require splenectomy but most will resolve with medical treatment.

3) Which of the following is NOT part of the immune system produced in the spleen?

a. IgG
b. IgM
c. Tuftsin
d. Properdin
e. Reticuloendothelial cells

Answer to question 3: (a): IgM, tuftsin, properdin, and reticuloendothelial cells are all produced in the spleen. The spleen does not produce IgG but is involved in producing some of the precursors. Splenectomized patients have a decrease the IgM immune globulin.

4) Which of the following is NOT associated with thrombotic thrombocytopenic purpura (TTP)?

a) Fever
b) Neurologic symptoms
c) Renal insufficiency
d) Decreased megakaryocytes in the bone marrow
e) Thrombocytopenia

Answer to Question 4: (d): The pentad of TTP is hemolytic anemia, purpura, thrombocytopenia, neurological symptoms, renal insufficiency in fever. There are normal or increased megakaryocytes as the body attempts to compensate for the thrombocytopenia.

## Further Reading

Blanchette V, Bolton-Maggs P. 2. Childhood immune thrombocytopenic purpura: diagnosis and management. Pediatr Clin North Am. 2008;55(2):393–420.

Chapman J, Bansal P, Goyal A, Azevedo AM. Splenomegaly. Stat Pearls (internet). 2021;11

Dubois L, Gray DK. Splenectomy: does it still play a role in the treatment of thrombocytopenic purpura? Can J Surg. 2010;53(5):349–55.

Grace RF, Zanella A, Neufeld EJ, Morton DJ, et al. Erythrocyte pyruvate kinase deficiency: 2015 status report. Am J Hematol. 2015;90:825–30.

Grace RF, Barcellini W. Management of pyruvate kinase deficiency in children and adults. Blood. 2019;136(11):1241–9.

Harrison’s principles of internal medicine. In: Disorders of Platelets and Vessel Wall, Longo, et al. editors, 18th edn. McGraw Hlll Pub.; 2012. p. 965–73.

Lambert MP, Gernsheimer TB. Clinical updates in adult immune thrombocytopenia. Blood. 2017;129:2929–35.

Sabiston textbook of surgery. In: The spleen, Townsend et al. editors, 21st ed. Elsevier Pub.; 2022. p. 1566–82.

Watts RG. Idiopathic thrombocytopenic purpura: a 10-year natural history study at the Children's Hospital of Alabama. Clin Pediatr (Philadelphia). 2004;43(8):691–702.

# Sickle Cell Disease

**Jeffrey Taylor**

**Key Points**

1) Sickle cell disease is a common inherited disease with numerous systemic manifestations.
2) Common procedures on sickle cell children requested of pediatric surgeons are central venous access, splenectomy, and cholecystectomy.
3) Surgical complications are common and close attention to perioperative care is necessary to avoid them.
4) Therapies including transfusions and hydroxyurea have improved the prognosis for many individuals with sickle cell disease who have access to care.
5) Most patients with sickle cell disease live in relative poverty and lack access to care which could prolong life and reduce symptoms.

1. Background and epidemiology:

   Sickle cell disease (SCD) is one of the most common inheritable diseases in the world with 30 million individuals affected [1]. The gene is prevalent in individuals from sub-Sahara Africa, the Indian subcontinent, the Mediterranean basin, the Middle East and in immigrants and their offspring from those areas [2]. In poor populations lacking state of the art medical care, it is a major cause of childhood morbidity and mortality with those under the age of 5 in Africa having a mortality rate as high as 50–90% [3]. The number of African adults

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with hemoglobin(hgb) SS is 20% of what would be expected based on newborn numbers [3].

The sickle gene and malaria occur in similar areas of the world and individuals with sickle cell trait (hgb A > S, 1 normal beta-globin and 1 abnormal beta-globin containing the sickle mutation) are protected from the severe forms of *Plasmodium falciparum* infection. Children at highest risk between 2 and 16 months of age have a 60% reduction in mortality if they possess sickle cell trait [4]. Kenyan children with sickle cell anemia (hgb SS) and severe *Plasmodium falciparum* infection had an 80% fatality rate compared to 10% in children with similar infection but who did not have SCD [5].

2. Pathophysiology:

   Sickle cell disease (SCD) results from the substitution of valine for glutamic acid at the 6th position of the beta-globin chain in hemoglobin (hgb). The substitution allows hgb S to polymerize in the de-oxygenated state and is what leads to the morbidity and mortality associated with vaso-occlusion and hemolysis. Factors which aid polymerization include stasis, inflammation, infection, increased blood viscosity, vasoconstriction, dehydration, cold, and acidosis. Up-regulation of adhesion molecules occurs which then leads to intravascular trapping of circulating sickled red blood cells, microvascular occlusion, and the activation of white blood cells and platelets which further stimulates deoxygenation and polymerization.

   SCD results from inheritance of at least 1 sickle beta-globin gene and 1 other abnormal beta-globin gene. In hgb SS, which is the most severe form, both beta-globins possess the S mutation; hgb S- beta thalassemia 0: 2nd beta-globin gene is non-functioning with no hgb A1 produced- similar in severity to hgb SS; hgb SC disease: 2nd beta-globin possesses the C mutation- usually but not always less severe than SS or S beta thal 0; hgb S-beta thalassemia+: 2nd beta-globin gene produces some normal beta-globin and thus some hgb A1 is produced, severity depends on the thalassemia genes output [6].

3. Clinical features:

   a. Vaso-occlusive pain (VOP):

      Sickled cells compared to normal red blood cells are less deformable, are more adherent to endothelial cells, and cause inflammation with increased coagulability which can result in vascular obstruction. This is accompanied with pain which may be severe [7]. Pain often begins in infancy with dactylitis and continues through life affecting the back, chest, abdomen, and extremities with many patients experiencing pain daily [7]. Importantly, the assessment of pain is by patient or parent report. No reliable laboratory testing or physical exam finding determines severity. In fact, the presence of abnormal vital signs, or changes in baseline lab values should alert the clinician that the patient is not experiencing a simple VOC pain episode and should prompt an investigation of complications such as acute chest syndrome (ACS), or splenic sequestration.

Pain may be associated with serious complications including ACS [8], multi-organ failure, defined as at least 2 of 3 lung, liver, or kidney failures [9], acute abdominal processes, transfusion reactions, spleen or liver sequestration, or avascular necrosis of bone such as the femur or humerus. Treatment of VOC pain is primarily opiate based although mild episodes may be relieved with Tylenol. Non-steroidal anti-inflammatory medications should be used with caution in patients with SCD due to their effects on the kidney.

b. Infection:

Children with SCD are at an increased risk for infection due to several factors including functional asplenia, hypoventilation/splinting from pain with resultant pneumonia/ACS, and from the presence of long-term catheters necessary for transfusions. Only 12% of children with hgb SS have normal splenic function by a mean age of 12 months [10]. Bacterial infections causing potential sepsis include *Streptococcus pneumoniae*, *Haemophilus influenza* type B, *E. coli*, *Staph aureus*, and Salmonella while *Mycoplasma pneumoniae* and Chlamydia as well as viruses cause pneumonia and ACS. Penicillin and vaccinations reduce but do not eliminate bacterial infection risk [11]. Certain viruses may cause significant problems in children with SCD such as red cell aplasia from Parvo B-19, and Covid-19 causing hypoxemia and a heightened inflammatory response leading to ACS [12, 13]. All children with hgb SS and hgb S- beta thal 0 should receive vaccinations including pneumococcal and H Flu type B as well as penicillin daily until age 5. Children with hgb SC or hgb S-beta thal + receive vaccinations with decisions concerning daily penicillin prophylaxis individualized.

Caregivers are educated concerning fever management and to seek medical care for a temperature of 101.5 F. or greater once, 100.4 F. or greater on 2 closely related occasions or ill appearance. Febrile children should receive a targeted but rapid physical exam, a CBC with differential, reticulocyte count and blood cultures [14] followed immediately by empiric parenteral antibiotics. Other testing as needed including urine, CXR, or lumbar puncture should be done where appropriate but should not delay antibiotics. Children requiring admission include those with associated complications such as ACS, VOC pain, splenic sequestration, or aplastic crisis; age < 2 with hgb SS or hgb S beta thal 0, temp >40 °C, WBC > 30,000 or <5000/mm$^3$, hgb <5 g/dl or decreased by 2 g compared to patient's baseline, history of previous invasive pneumococcal disease, presence of an indwelling catheter, ill appearance, allergy to long acting ceftriaxone and thus receiving clindamycin or vancomycin, or concerns about a family's risk for lack of follow up [15, 16].

c. Anemia:

Hemolysis resulting in anemia is a hallmark of SCD and is chronic, but patients may experience acute exacerbations. Besides baseline chronic hemolysis with shortened red blood cell life span, patients may have iron deficiency, and/or a decreased erythropoietin drive from sickle cell related kidney disease [17].

Acute decreases in hemoglobin may occur with aplastic crisis, splenic sequestration, hyper-hemolytic crisis or may be an early presenting sign of impending ACS. Sickle cells have a typical lifespan of 10–17 days as opposed to 120 days for normal red blood cells [18]. Typical hemoglobin values in patients with SCD are 8–10 g/dl but may be lower and it is important to know an individual patient's baseline level for treatment decisions. Baseline reticulocyte counts are elevated consistent with shortened RBC lifespan, and the white blood count is often elevated. Consistent with hemolysis, LDH, and unconjugated bilirubin are often elevated [19].

Severe anemia secondary to aplastic crises occurs due to the shortened RBC lifespan of sickle cells. Splenic sequestration occurs secondary to the trapping of red blood cells in the spleen and may be rapid. Children may present in shock which can be fatal. Young children with hgb SS or hgb S beta thal 0 are affected prior to their spleens becoming fibrotic, while individuals with milder forms of disease present later in life [20]. Patients present with a rapidly enlarging spleen, low hgb, and often a brisk reticulocytosis. Conversely, aplastic crisis, unless caught in the recovery period, usually has a low reticulocyte count. Prompt fluid resuscitation and packed red blood cells are necessary and lifesaving. Many (78%) will have a second sequestration event [21]. Severe or recurrent episodes require splenectomy.

d. Neurologic

Both ischemic and hemorrhagic strokes occur in patients with SCD. Prior to the use of trans-cranial Doppler, 11% of children experienced a clinically apparent stroke by age 20. Transient ischemic attacks (TIAs) and silent cerebral infarcts (SCIs) also occur and often result in neurocognitive and behavioral deficits. Prevention of stroke is of the utmost importance and transcranial Doppler (TCD) is used to screen for stroke risk beginning at age 2. Children at risk by TCD undergo transfusion therapy for prevention [22] with a goal of keeping hgb S < 30%. Most patients receive transfusions indefinitely, although a subset may be switched to hydroxyurea [23, 24] or undergo stem cell transplant with a matched sibling donor [23, 25]. TCD is not effective in screening for silent cerebral infarcts, and children with hgb SS or hgb S beta thal 0 should have at least 1 MRI without sedation during school age or young adulthood [23]. Transfusion therapy is necessary to prevent stroke recurrence (24).22 Consideration of stem cell transplant in select patients unable to receive transfusions may also be a viable alternative.

e. Pulmonary

Pulmonary arteries have low PO2, and lower flow compared to systemic circulation which promotes sickling. Complications include ACS, asthma, pulmonary fibrosis, and pulmonary hypertension. ACS is a leading cause of death in SCD. 50% of individuals who develop ACS while hospitalized were admitted for a different reason. Many of these patients will require mechanical ventilation. Only 38% have an identifiable cause [26].

Patients with ACS present with fever, hypoxemia, cough, wheezing, respiratory distress, pain, and infiltrates on CXR. 50% of patients with SCD will experience an ACS which is a major cause of morbidity and mortality. Causes are varied and in addition to those listed above include vaso-occlusive pain, hypoventilation which may be secondary to pain, atelectasis, and thrombosis/thromboembolism. Severe pain leads to splinting, hypoventilation, and hypoxia. Long-term complications are common and include pulmonary hypertension and pulmonary insufficiency.

Management includes careful use of analgesia, oxygen, ventilatory support when needed, incentive spirometry—used both therapeutically after ACS has occurred and as prevention in patients admitted for other reasons, bronchodilators, antibiotics to include macrolides, judicious use of fluids, and transfusion support [26]. Exchange transfusion should be reserved for progressive disease, severe hypoxia, or multiple lobe involvement. Inhaled nitric oxide is currently being studied and extracorporeal membrane oxygenation (ECMO) may be considered for patients not responding to mechanical ventilation. The role of steroids is questionable due to the risk of rebound ACS when stopped.

Asthma is more common is children with SCD and is a risk factor for ACS. Additionally, patients with a history of ACS are at increased risk for asthma [27, 28]. Asthma should be aggressively treated in children with SCD.

Sleep disordered breathing and nocturnal hypoxemia occur in as many as 79% of children with SCD [29] and is superimposed on an already decreased oxygen carrying capacity. These children have increased risk of vaso-occlusion and neurologic complications and correction of sleep disordered breathing decreases vaso-occlusion, ACS, and cerebrovascular problems [30].

Pulmonary hypertension occurs in 6–10% of patients with sickle cell disease [31, 32]. These patients may have vague symptoms such as reduced exercise tolerance, dyspnea, or presyncope. There is controversy regarding whether screening with transthoracic echocardiogram in asymptomatic patients is necessary [33, 34] with some suggesting beginning at age 8 while others wait until adolescence.

f. Renal

Twenty-five percent of older individuals with sickle cell disease will develop renal insufficiency [35]. The cause of kidney disease is multifactorial. Early in life glomerular hyperfiltration occurs with kidney hypertrophy and urinary concentrating ability is lost. Creatinine is generally normal [36]. Proximal tubule function may be increased, and the resultant elevated glomerular filtration rate (GFR) may be misleading. This is followed by microalbuminuria and hematuria, a decrease in GFR, and rise in creatinine. Some individuals will progress to focal segmental glomerulosclerosis which leads to end stage kidney disease [36]. Factors contributing to renal damage include papillary necrosis, infarction, medication toxicity including non-steroidal anti-inflammatory medications, and hypertension. Nephrotoxic drugs should be avoided or limited. Hypertension should be aggressively treated. A rare but serious malignancy, renal medullary carcinoma, occurs in patients with sickle cell trait and less often in other sickle syndromes. New flank pain or hematuria should prompt investigation [37].

g. Bone

Children with sickle cell disease suffer from bone complications due to vaso-occlusion, osteonecrosis, and infections. Bones containing red marrow are at risk. Vertebral body collapse and avascular necrosis of bones such as the femoral heads are significant complications. Dactylitis is common early in life with 45% of children with sickle cell disease experiencing at least 1 episode by age 2 [38].

A difficult problem is differentiating osteomyelitis and septic arthritis from the much more common vaso-occlusive bone pain. Many patients can differentiate their "typical" vaso-occlusive pain from that of osteomyelitis and may be able to help in their own diagnosis. Patients with osteomyelitis have a higher CRP and temperature than those that do not. White blood counts are similar, and pain improves without antibiotics in patients with vaso-occlusive pain [39]. Bacterial organisms causing osteomyelitis include Salmonella, *E. coli* and *S. aureus* [40].

Avascular necrosis (AVN) of the femoral (and humeral) heads are complications of SCD. AVN is progressive over time with many patients going on to joint destruction. Pain can become debilitating with many requiring surgery/joint replacements. Bone marrow expansion compensating for hemolysis causes widened medullary spaces and osteoporosis. Many children are vitamin D deficient [41] requiring replacement.

h. Cardiac

Cardiomyopathy with left diastolic dysfunction is common [42], and is associated with pulmonary hypertension, chronic anemia with hypoxemia, increased cardiac output, dilated left ventricle, iron overload, hypertension, and renal insufficiency [43].

i. Hepatobiliary

Children with sickle cell disease are at risk for liver toxicity. Liver failure is associated with ischemia, cholestasis, iron overload, sequestration events, gallstones, and viral infections including hepatitis C. Hemolysis leads to increased bile acids resulting in pigmented gallstones. Children with sickle cell disease are at increased risk for symptomatic cholelithiasis and cholecystitis.

j. Skin

Lower leg ulcers are common over age 10, and the risk of infection is high. The presence of a new leg ulcer should prompt an evaluation for deep vein thrombosis [44].

k. Eye

Retinopathy with retinal artery occlusion, ischemia, and proliferative retinopathy may begin in adolescence and is more common in hgb SC disease [45].

l. Priapism

Priapism is a relatively common problem and if persisting for greater than 4–6 h is a urologic emergency to prevent erectile dysfunction. Most priapism in SCD is low flow ischemic priapism. It behaves as a compartment syndrome with decreased or absent cavernous blood flow. Acute management includes alpha-adrenergic medications such as phenylephrine and aspiration of blood from the corpus cavernosum [46]. Surgical management is occasionally needed.

4. Surgical Considerations and Transfusion

   Patients with SCD have increased morbidity perioperatively including vaso-occlusive crises, ACS, infections, heart failure, strokes, renal complications, and mortality. Common surgical procedures in sickle cell patients are central venous access, cholecystectomy, splenectomy, tonsillectomy, umbilical hernia repair, orthopedic procedures, and appendectomy. Pre-operative evaluation is helped if SCD is looked upon as a systemic disease with the potential to affect all organs. The type of sickle cell (SS, SC, S beta thalassemia), baseline hematologic parameters including hemoglobin, hematocrit, reticulocyte count, white blood cell count, platelets, crises history, opioid use, and transfusion history including presence of antibodies which would delay blood availability are important in the preoperative evaluation of the sickle cell patient. Prolonged pre-op fluid restriction should be avoided when possible since dehydration is a risk factor for polymerization. Choice of fluids must take sickle cell risks into account. Normal saline should be used only for deficit replacement as it is acidic and increases viscosity. Relatively hypotonic fluids guided by electrolyte measurements, are preferred for maintenance. Hypoxia must be avoided, and patients kept warm. Post-op use of incentive spirometry decreases atelectasis and ACS [1]. Although some debate exists [47], the practice in the USA is to use simple transfusions preoperatively to bring the hemoglobin to 10 g/dl in patients undergoing general anesthesia lasting more than one hour [48] with exchange transfusions to bring hgb S to <30% of total (or <5% for cardiopulmonary bypass) considered for high-risk surgery, and in patients with severe sickle cell disease and/or significant co-morbidities [47, 49]. Minor procedures in patients such as skin biopsy or myringotomy tube placement may do well without preoperative transfusion. Blood should be leukocyte reduced and sickle cell trait blood should not be used.

   Laparoscopic procedures are now routinely used in many centers with reduced post-op complication rates. Pain control in sickle cell patients with prior exposure to opiates necessitates higher than standard doses. Strict use of incentive spirometry, especially with opioid medications, is helpful in decreasing post-operative ACS. The use of deep vein thrombosis prophylaxis, while used in adults, is not routinely used in children unless previous thrombosis or other significant risk factors are present [1].

5. Sickle Cell Trait

   Sickle Cell Trait (SCT) is generally benign with an incidence in African American newborns of 73/1000 and 7/1000 in Hispanics. Infants are identified via newborn screening which occurs in all states. Counseling of these individuals is especially important prior to childbearing and intensive physical training. In the US 2.5 million individuals have SCT (hgb A > S) with 300 million globally [50].

   Patients with SCT lose the ability to concentrate urine as the hypertonic conditions in the renal medulla favor sickling even in those with trait [51]. These children may display nocturnal enuresis and are at increased risk for dehydration. Patients are at increased risk for proteinuria, hematuria, and chronic kidney disease [51, 52]. Patients with SCT and hematuria must also be evaluated for renal medullary carcinoma.

Individuals with SCT are at increased risk for venous thromboembolism and pulmonary embolism, the risk approximates the risk level of patients heterozygous for the prothrombin gene mutation or factor V Leiden [52]. There is no increase in cardiovascular risk, heart failure, fitness, metabolic syndrome, diabetes, diabetic or non-diabetic end stage renal disease or mortality occurs in patients with SCT [51, 52].

## Additional Notes

Preventive and Curative Strategies in Patients with SCD:

Hydroxyurea, an anti-cancer drug, was approved for use in SCD in 1998. It inhibits the production of red blood cells, platelets, and white blood cells. Its value in SCD is due to its effect on gamma globin production which it increases resulting in an overall increase in fetal hemoglobin. Increasing hgb F compared to hgb S delays the polymerization of hgb S, increases red blood cell lifespan, decreases adhesion to endothelium and increases cellular hydration [53, 54]. Hemoglobin F levels >20% have a beneficial effect on vaso-occlusion, hospitalization, ACS, and fever [55]. Hydroxyurea is offered to all patients with hgb SS and S beta thal 0 beginning at age 9 months through adolescence and to patients with milder forms if severe symptoms are present. Hydroxyurea use in infants resulted in less pain, dactylitis, ACS, gastroenteritis, transfusions, and a trend toward improved splenic function [56, 57].

Hydroxyurea has no role in the management of acute episodes. Side effects including pancreatitis, worsening kidney function, or severe reactions require dose reductions or cessation of therapy.

Stem cell transplant is a potential treatment option for select children with SCD resistant to other treatments and is the only curative therapy available. Several issues regarding transplant remain including lack of randomized controlled trials comparing transplant to medical therapy. At this time transplant may be an option for select patients but more research needs to be done before becoming widely available [33].

## Study Questions

1) Surgical preparation for a patient with Sickle Cell Disease includes all the following except:

   a) Transfusion prior to the surgery to at least 12 g/dl so that the patient will have as much oxygen carrying capacity as possible

   b) Discuss with the patient or caretakers what the usual opiate need is during vaso-occlusive crises
   c) Stressing with the patient and family the use of incentive spirometry post-op

Answer: (a) is correct answer. Hemoglobin greater than 10 g/dl is associated with increased blood viscosity which favors polymerization.

(b) would be appropriate for surgical preparation—patients with sickle cell disease may have an extensive history of opiate exposure. Parents are often the best resource for appropriate dosing. (c) would be appropriate for surgical preparation—incentive spirometry is critical in preventing ACS post operatively. Younger children may be more likely to use spirometry if you have enlisted parental support.

2) Sickle Cell Trait is associated with:

   a) Increased surgical complications
   b) Increased risk of stroke in the immediate post-op period
   c) Loss of urine concentrating ability making fluid management more difficult

Answer: (c) The hypertonicity of the renal medulla favors sickling even in patients with SCT. This damage leads to the inability of children with SCT to concentrate their urine thus rendering urine specific gravity less useful in deciding on a patient's hydration status (a) and (b) are not true

## References

1. Adjepong KO, Otegbeye F, Adjepong YA. Perioperative management of sickle cell disease. Mediterr J Hematol Infect Dis. 2018;10:e2018032.
2. Piel FB, Patil AP, Howes RE, Nyangiri OA, Gething PW, Dewi M, Temperley WH, Williams TN, Weatherall DJ, Hay SI. Global epidemiology of sickle haemoglobin in neonates: a contemporary geostatistical model-based map and population estimates. Lancet. 2013;381:142–51.
3. Grusse SD, Odame I, Atrash HK, Amendah DD, PielFB WTN. Sickle cell disease in africa: a neglected cause of early childhood mortality. Am J Prev Med. 2011;41(Suppl 4):S398–405.
4. Aidoo M, Terlouw D, Kolczak M, McElroy P, Kuile F, Kariuki S, et al. Protective effects of the sickle cell gene against malaria morbidity and mortality. Lancet. 2002;359:1311-1312
5. McAuley CF, Webb C, Makani J, et al. High mortality from plasmodium falciparum malaria in children living with sickle cell anemia on the Coast of Kenya. Blood. 2010;116:1663.
6. Rees DC, Williams TN, Gladwin MT. Sickle cell disease. Lancet. 2010;376:2018.
7. Platt OS, Therington BD, Brambilla DJ, et al. Pain in sickle cell disease. Rate and risk factors. N Engl J Med. 1991;325(11)
8. Vichinsky EP, Styles LA, Colangelo LH, et al. Acute chest syndrome in sickle cell disease: clinical presentation and course. Cooperative study of sickle cell disease. Blood. 1997;89:1787–92.
9. Hassel KL, Eckman JR, Lane PA. Acute multi organ failure syndrome: a potentially catastrophic complication of severe sickle cell pain episodes. Am J Med. 1994;96:155.
10. Rogers ZR, Wang WC, Luo Z, et al. Biomarkers of splenic function in infants with sickle cell anemia: baseline data from the BABY HUG trial. Blood. 2011;117:2614.
11. Quinn CT, Rogers ZR, Buchanan GR. Survival of children with sickle cell disease. Blood. 2004;103:4023.

12. Nur E, Gaartman AE, van Tuijn CFJ, et al. Vaso-occlusive crisis and acute chest syndrome in sickle cell disease due to 2019 novel coronavirus disease (COVID-19). Am J Hematol. 2020;95:725.
13. McCloskey KA, Meenam J, Hall R, Tsitsikas DA. COVID-19 infection and sickle cell disease: a UK centre experience. Br J Haematol. 2020;190:e57.
14. Wang CJ, Kavanagh PL, Little AA, et al. Quality of care indicators for children with sickle cell disease. Pediatrics. 2011;128:484.
15. Mountain States Regional Genetic Services Network. Sickle cell disease in children and adolescents: diagnosis, guidelines, for comprehensive care and protocols for management of acute and chronic complications. 2001
16. Wilmas JA, Flynn PM, Harris S, et al. A randomized study of outpatient treatment with ceftriaxone for selected febrile children with sickle cell disease. N Engl J Med. 1993;329:472.
17. Sherwood JB, Goldwasser E, Chilcote R, et al. Sickle cell anemia patients have low erythropoietin levels for their degree of anemia. Blood. 1986;67:46.
18. McCurdy PR, Sherman AS. Irreversibly sickled cells and red cell survival in sickle cell anemia: a study with both DF32P and 51CR. Am J Medic. 1978;64(2):253.
19. West MS, Wethers D, Smith J, Steinberg M. Laboratory profile of sickle cell disease: a cross sectional analysis. The cooperative study of sickle cell disease. J Clin Epidemiol. 1992;45:893.
20. Emond AM, Collis R, Darvill D, et al. Acute splenic sequestration in homozygous sickle cell disease: natural history and management. J Pediatr. 1985;107:201.
21. Ben Khaled M, Ouederni M, Mankai Y, et al. Prevalence and predictive factors of splenic sequestration crisis among 423 pediatric patients with sickle cell disease in Tunisia. Blood Cells Mol Dis. 2020;80:102374.
22. Kassim AA, Galadanci NA, Pruthi S, DeBaun MR. How I treat and manage strokes in sickle cell disease. Blood. 2015;125:3401.
23. DeBaum MR, Jordan LC, King AA, et al. American society of hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults. Blood Adv. 2020;4:1554.
24. Ware RE, Davis BR, Schultz WH, et al. Hydroxycarbamide versus chronic transfusion for maintenance of transcranial doppler flow velocities in children with sickle cell anemia- TCD with transfusion changing to hydroxyurea (TWITCH): a multicentre open-label, phase 3. Non-inferiority trial. Lancet. 2016;387:661.
25. Bernaudin F, Verlhae S, Peffault de Latour R, et al. Association of matched sibling donor hematopoetic stem cell transplantation with trans cranial Doppler velocities in children with sickle cell anemia. JJAMA. 2019;321:266.
26. Vichinsky EP, Neumeyr LD, Earles AN, et al. Causes and outcomes of the acute chest syndrome in sickle cell disease. National acute chest syndrome study group. N Engl J Med. 2000;342:1855.
27. Mehari A, Klings ES. Chronic pulmonary complications of sickle cell disease. Chest. 2016;149:1313.
28. DeBaum MR, Strunk RC. The intersection between asthma and acute chest syndrome in children with sickle cell anaemia. Lancet. 2016;387:2545.
29. Kaleyais J, Mostofi N, Grant M, Colman C, et al. Severity of obstructive sleep apnea in children with sickle cell disease. J Pediatr Hematol Oncol. 2008;30:659.
30. Tripathi A, Jerrell JM, Stallworth JR. Cost-effectiveness of adenotonsillectomy in reducing obstructive sleep apnea, cerebrovascular ischemia, vaso-occlusive pain, and ACS episodes in pediatric sickle cell disease. Ann Hematol. 2011;90:145.
31. Hawkins J, Jeng M, Harris S, et al. Chronic transfusion therapy for children with sickle cell disease and recurrent acute chest syndrome. J Pediatr Hematol Oncol. 2005;27:158.
32. Parent F, Bacher D, Inamo J, et al. A hemodynamic study of pulmonary hypertension in sickle cell disease. N Engl J Med. 2011;365:44.
33. Yawn BP, Buchanan GR, Afeny-Annan AM, et al. Management of sickle cell disease: summary of the 2014 evidence-based report by expert panel members. JAMA. 2014;312:1033.

34. Liem RI, Lanzkron S, Coates T, et al. American society of hematology 2019 guidelines for sickle cell disease: cardiopulmonary and kidney disease. Blood Adv. 2019;3:3867.
35. Vichinsky E. Chronic organ failure in adult sickle cell disease. Hematol Am Soc Hematol Educ Prog. 2017;2017:435.
36. Wesson DE. The initiation and progression of sickle cell nephropathy. Kidney Int. 2002;61:2277.
37. Alvarey O, Rodriquez MM, Jordan L, Sarnack S. Renal medullary carcinoma and sickle cell trait: a systematic review. Pediatric Blood Cancer. 2015;62:1694.
38. Gill FM, Sleeper LA, Weiner SJ, et al. Clinical events in the first decade in a cohort of infants with sickle cell disease. cooperative study of sickle cell disease. Blood. 86:776.
39. Fontalis A, Hughes K, Nguyen MP, et al. The challenge of differentiating vaso-occlusive crises from osteomyelitis in children with sickle cell disease and bone pain: a 15-year retrospective review. J Child Orthop. 2019;13:33.
40. Kaplan J, Ikeda S, McNeil JC, et al. Microbiology of osteoarticular infections in patients with sickle hemoglobinopathies at Texas Children's Hospital, 2000-2018. Pediatr Infect Dis J. 2019;38:1251.
41. Lal A, Fung EB, Pakbaz Z, et al. Bone mineral density in children with sickle cell anemia. Pediatr Blood Cancer. 2006;47:901.
42. Gladwin MT, Sachdev V. Cardiovascular abnormalities in sickle cell disease. J Am Coll Cardiol. 2012;59:1123.
43. Zilberman MV, Du W, Das S, Sarnaik SA. Evaluation of left ventricular diastolic function in pediatric sickle cell disease patients. Am J Hematol. 2007;82:433.
44. Minniti CP, Delaney KM, Gorbach AM, et al. Vasculopathy, inflammation, and blood flow in leg ulcers of patients with sickle cell anemia. Am J Hematol. 2014;89:1.
45. Downes SM, Hambleton IR, Chuang EL, et al. Incidence and natural history of proliferative sickle cell retinopathy: observations from a cohort study. Ophthalmology. 2005;112:1869.
46. Anele VA, Le BV, Resar LM, Burnett AL. How I treat priapism. Blood. 2015;125:3551.
47. Schyrr F, Dolci M, Nydegger M. Perioperative care of children with sickle cell disease: a systematic review and clinical recommendations. Am J of Hematol. 2019;95:78.
48. Chou ST, Alsawas M, Fasano RM, et al. American society of hematology 2020 guidelines for sickle cell disease: transfusion support. Blood Adv. 2002;4:327.
49. Davis BA, Allard S, Qureshi A, et al. Guidelines on red cell transfusion in sickle cell disease Part II: indications for transfusion. Br J Haematol. 2017;176:192.
50. O'Connor FG, Bergeron MF, Cantrell J, et al. ACSM and CHAMP summit on sickle cell trait: mitigating risk for war fighters and athletes. Med Sci Sports Exerc. 2012;44:2045.
51. Xu JZ, Thein SL. The carrier state for sickle cell disease is not completely harmless. Haematologica. 2019;104:1106.
52. Naik RP, Smith-Whitley K, Hassel KL, et al. Clinical outcomes associated with sickle cell trait: a systematic review. Ann Intern Med. 2018;169:619.
53. Franco RS, Yasin Z, Palascak MB, et al. The effect of fetal hemoglobin on the survival characteristics of sickle cells. Blood. 2006;108:1073.
54. Setty BN, Kulkarni S, Dampier CD, Stuart MJ. Fetal hemoglobin in sickle cell anemia: relationship to erythrocyte adhesion markers and adhesion. Blood. 2001;97:2568.
55. Estepp JH, Smeltzer MP, Kang G, et al. A clinically meaningful fetal hemoglobin threshold for children with sickle cell anemia during hydroxyurea therapy. Am J Hematol. 2017;92:1333.
56. Wang WC, Ware RE, Miller ST, et al. Hydroxycarbamide in very young children with sickle cell anemia: a multicentre, randomized controlled trial (BABY HUG). Lancet. 2011;377:1663.
57. Thornburg CD, Files BA, Luo Z, et al. Impact of hydroxyurea on clinical events in the BABY HUG trial. Blood. 2012;120:4304.

# Empyema and Thoracoscopic Drainage

**Tiffany Zens, Kara Gill, and Daniel Ostlie**

**Key Points**

1) Although bacterial pneumonia is decreasing in the pediatric population in the USA, the rate of infectious complications of pneumonia is increasing.
2) Most cases of empyema can be managed non-operatively with fibrinolytic agents.
3) Thoracoscopic drainage for empyema should be reserved for patients that fail fibrinolytic treatment.

1. Epidemiology:

    a. Incidence: The rates of pediatric bacterial pneumonia have been declining over the last several decades, accounting for approximately 3 million outpatient visits and 150,000 hospitalizations in the United States annually [1–4]. This decrease is likely a result of the development of the pneumococcal conjugate vaccine for community-acquired pneumonia [1, 5]. Despite the decrease in pediatric pneumonia, the rates of infectious complications like parapneumonic effusion and empyema are increasing. Empyema occurs in

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0.6–2% of all pediatric pneumonias and 2–8% of hospitalized pediatric pneumonias [6–12]. Furthermore, 35–58% of empyema patients will require ICU level care for management of their pulmonary disease [13, 14].

b. Etiology: Studies found that recent varicella-zoster viral infection increases a child's risk for developing empyema, but patient age, patient gender, maternal age, parental smoking, socioeconomic status and asthma were not risk factors [15]. *Streptococcus pneumoniae*, followed by *Staphylococcus aureus* and *Streptococcus pyogenes* are the most common bacterial pathogens.

2. Pathophysiology:

   a. The pathophysiology of pediatric empyema is not completely understood, but it is thought to be a progressive disease that occurs in stages. In the initial exudative stage, pleural and parenchymal inflammation result in increased capillary permeability and fluid production. This results in an uncomplicated parapneumonic effusion in which the fluid is free flowing with a low pleural white blood cell (WBC) count. In the second fibrinopurulent stage, infected pleural fluid stimulates cytokine release and results in activation of the coagulation cascade, production of fibrin and purulence. Fibrin strands form loculations and septations which block lymphatic drainage, causing further accumulation of intrathoracic fluid. Finally, in the organizing stage, increased fibroblast production causes thickening of the pleura. An inflammatory rind or peel forms, which can limit the ability of the lung to expand, resulting in a trapped lung [6, 16, 17].

3. Diagnosis:

   a. *History and Physical*: Pediatric empyema is diagnosed through careful review of the child's history and physical exam, imaging findings and laboratory results. A child with empyema often presents with a history of a recent thoracic infectious process or thoracic trauma. Symptoms at presentation include fevers, shortness of breath, hypoxia, tachypnea, chest pain and cough. On exam, children exhibit decreased lung sounds on the affected side, dullness to percussion, and in severe cases, increased respiratory effort.
   b. *Imaging*:

      i. *Plain Film Radiographic Imaging:* A chest X-ray (CXR) is often the initial test performed to evaluate for thoracic disease in children. In the case of empyema, CXR will often demonstrate obliteration of the costophrenic angle with a layer of fluid tracking up the lateral chest wall. A pleural cap is present if fluid accumulates at the apex of the lung. Additional upright or decubitus views can help differentiate whether fluid is free-flowing or loculated. In severe cases, CXR can reveal complete opacification of the affected hemithorax or contralateral displacement of the mediastinal structures (Fig. 1). Unfortunately, it is often difficult to distinguish whether opacities on CXR are a result of parenchymal disease or pleural

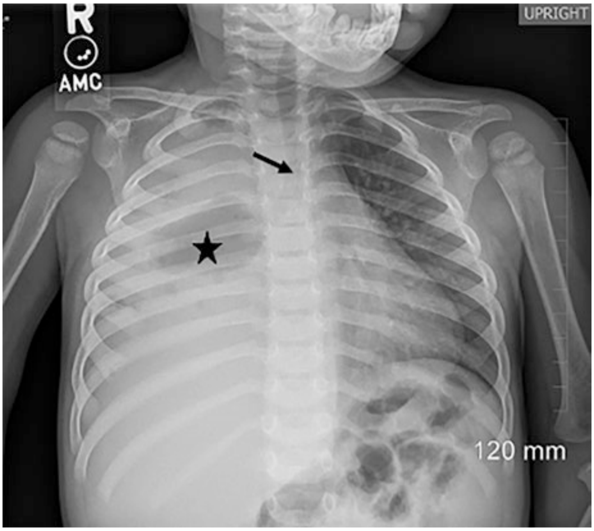

**Fig. 1** CXR Empyema: Upright AP chest radiograph in a 14-month-old male shows near diffuse opacification of the right hemithorax with leftward shift of the mediastinum (arrow) which can be seen with pneumonia complicated by effusion or empyema. The ovoid lucency (star) in the central right lung is concerning for a pneumatocele vs. loculated air in the pleural space

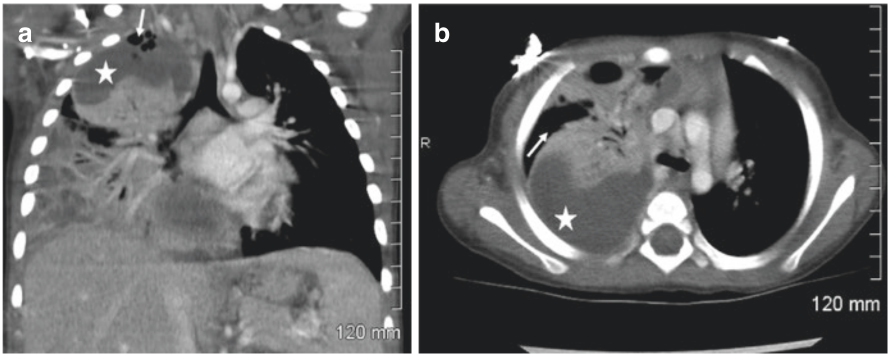

**Fig. 2** CT Empyema: Contrast enhanced coronal reformatted (**a**) and axial (**b**) images of the chest shows diffuse opacification of the right lung with areas of nonenhancement consistent with necrotizing pneumonia. Fluid within the pleural space over the lung apex (stars) exerts mass effect on the right upper lobe causing it to have a scalloped margin, consistent with empyema. There is also air in the pleural space (arrows) due to the presence of a bronchopleural fistula

disease. As a result, additional imaging is almost always needed for the diagnosis and treatment of empyema [9].

ii. *Cross-sectional Imaging:* Computer Tomography (CT) allows for a cross-sectional evaluation of the entire thoracic cavity. Figure 2 demonstrates CT findings consistent with empyema, concomitant necrotizing pneumonia, and broncho-pleural fistula. Given the risk of cancer associated with ionizing radiation in children, pediatric dosing strategies are recommended for the evaluation of pediatric empyema. CT examinations should be done with IV contrast to allow pleural enhancement and help differentiate between pleural and parenchymal disease. CT is most useful for complicated cases involving necrotizing pneumonias with cavitation or abscesses, endobronchial obstructions, mediastinal disease, or broncho-pleural fistulas [9].

iii. *Sonographic Imaging:* Ultrasound (US) has become the gold standard imaging modality for the evaluation of empyema and parapneumonic effusions in children. US allows dynamic imaging of the pleural space without radiation exposure. It can identify increased cellularity and debris, hemorrhage, fibrin deposits and loculations within an empyema or effusion. Furthermore, US can easily distinguish between an avascular pleural fluid collection and a vascular parenchymal lung consolidation. US is highly sensitive and can detect even small effusions. Dynamic US imaging can determine if pleural fluid is freely moving or loculated. Additionally, dynamic images can evaluate for adequate lung expansion or evidence of trapped lung. Finally, US can be used therapeutically, to help guide proper chest tube placement and ensure optimum drainage. Figure 3 shows common US findings of empyema. With the exception of complicated cases involving lung abscesses or broncho-pleural fistulas, studies have shown no significant benefit of CT over US imaging for the evaluation of empyema [9, 18, 19]. As a result, the American Pediatric Surgical Association (APSA) Outcomes and Clinical Trials Committee has recommended US as the primary imaging modality for the evaluation of pediatric empyema [6].

c. *Laboratory Analysis*: Standard laboratory testing for patients with empyema, particularly those who are critically ill, include plasma complete blood count, ESR/CRP, coagulation panel, and chemistry panel. Blood cultures should be obtained, particularly in patients who are febrile. Studies demonstrate positive blood cultures in 7–23% of children with empyema [7, 13, 14, 20, 21].

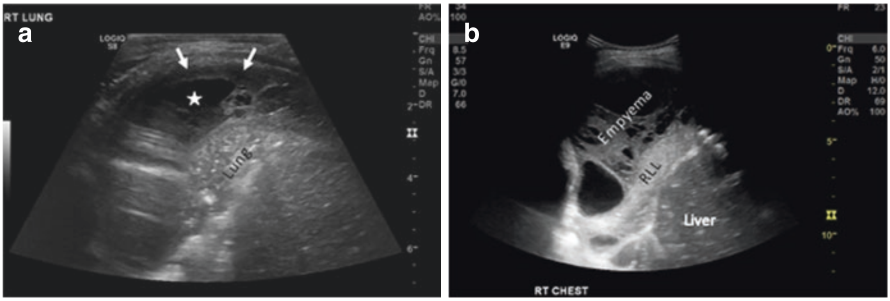

**Fig. 3** US Empyema: (**a**) Grayscale sonographic images of the chest in a 17 month-old male reveals thickened pleura (arrows) as well as heterogenous echogenicity of the pleural fluid with thick septations (star). The lung underlying this empyema is more hypoechoic than normal, consistent with pneumonia. (**b**) Gray-scale sonographic image of the lower right chest in a 6 year-old female reveals complex fluid with thick septations (star) causing mass effect on the underlying right lower lobe (RLL), consistent with empyema

Additionally, pleural fluid should be sampled and sent for evaluation. Empyema is suspected when pleural fluid demonstrates a WBC > 10,000 $\text{mm}^3$ with a neutrophil predominance, glucose <40 mg/dL or half serum glucose level, high protein, lactate dehydrogenase (LDH) greater than 1000 U/L or three times the upper limit for serum LDH, and a pH < 7.2 [6, 22–24]. Pleural fluid should also be sent for gram stain and culture to help identify infectious organisms and aid in antibiotic selection. Pleural fluid cultures can often be sterile secondary to prolonged antibiotic administration prior to sampling. Studies have demonstrated that approximately 30–35% of empyema pleural cultures grow the causative bacteria; however, pathogen identification can be improved with the use of PCR [10, 20, 21]. The most common organism found in pleural fluid is *Streptococcus pneumoniae*, followed by *Staphylococcus Aureus* and *Streptococcus pyogenes,* and, less commonly, *Haemophilus influenzae, Enterobacteriaceae,* anaerobes, *Legionella, Histoplasma, Coccidioides, Aspergillus, Entamoeba, Nocardia,* and *Mycobacterium tuberculosis* [22, 25, 26]. If the patient has risk factors for pleural tuberculosis, special acid-fast testing should be sent.

4. Treatment: Historically, there has been significant controversy regarding the treatment of pediatric empyema and the role of surgery. This controversy was a result of several retrospective studies demonstrating a benefit to early operative intervention and questioning the efficacy of conservative management [12, 27, 28]. Recently, with the addition of intrathoracic fibrinolytic therapies, three landmark randomized controlled trials have demonstrated equivalent outcomes with medical and surgical approaches. As a result, The American Association for Thoracic Surgery, British Thoracic Society, European Thoracic Surgery Guidelines, and APSA all advocate for a similar stepwise approach to the treatment of pediatric empyema [6, 17, 29, 30]. The APSA treatment algorithm for empyema is showed in Fig. 4. Once an empyema has been identified, a small-bore chest tube is inserted. The patient is then treated with daily intrathoracic doses of tissue plasminogen activator (tPA) for 3 days (4 mg tPA/40ml saline with a 1-hour dwell). If the patient does not clinically improve, the child is re-imaged. If persistent pleural disease is identified, the child undergoes Video-Assisted Thoracoscopic Surgery (VATS) for drainage and possible decortication. If no pleural disease is present, the child is treated with antibiotics and medical management.

   a. *Medical Treatment*:

      i. *Drainage and Source Control:* Pediatric empyema is initially treated with drainage and source control of the infected pleural fluid. Unlike other pediatric effusions, simple thoracentesis is rarely indicated for empyema outside very early-stage disease with free-flowing fluid. Paracentesis does not allow for continuous drainage or instillation of intrathoracic medications. The APSA Outcomes and Clinical Trials Committee has recommended against repeat thoracentesis in the treatment of pediatric

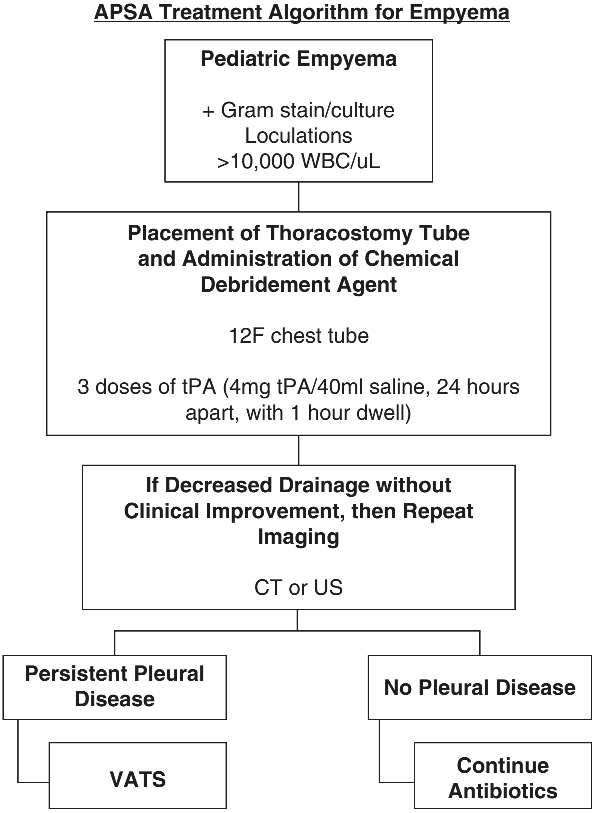
APSA Treatment Algorithm for Empyema
Pediatric Empyema
+ Gram stain/culture
Loculations
>10,000 WBC/uL
Placement of Thoracostomy Tube and Administration of Chemical Debridement Agent
12F chest tube
3 doses of tPA (4mg tPA/40ml saline, 24 hours apart, with 1 hour dwell)
If Decreased Drainage without Clinical Improvement, then Repeat Imaging
CT or US
Persistent Pleural Disease
No Pleural Disease
VATS
Continue Antibiotics

**Fig. 4** This algorithm was obtained from APSA Outcomes and Clinical Trials Committee recommendations. Islam S, Calkins CM, Goldin AB, et al. The diagnosis and management of empyema in children: a comprehensive review from the APSA Outcomes and Clinical Trials Committee. J Pediatr Surg. Nov 2012;47(11):2101-10

empyema [6]. Instead, experts advocate for chest tube placement at the bedside or with ultrasound guidance under sedation. Studies have demonstrated that there is no difference in the ability to drain pleural fluid or administer fibrinolytic therapy using a smaller pigtail chest tube compared to large-bore chest tubes [31]. Therefore, to limit patient discomfort, small chest tubes (12–14F) should be placed for drainage of empyema.

ii. *Antibiotics:* Antibiotics should be guided by the patient's culture results, age, vaccine status and community antibiograms. In most cases, patients are initially treated with a third-generation cephalosporin plus an anti-staphylococcal beta-lactamase resistant penicillin. Additionally, Clindamycin can be added for *Pneumococci* or anaerobic coverage, or vancomycin can be added if *Methicillin-Resistant Staphylococcus aureus* is suspected [22]. Although there is no consensus on the exact duration of the antibiotic course, the APSA Outcomes and Clinical Trials Committee has recommended a minimum of 10 days of antibiotic therapy after resolution of fevers [6].

iii. *Intrapleural Administration of Fibrinolytics:* Intrathoracic fibrinolytic agents work by degrading fibrin within the empyema, thus allowing drainage of inflammatory debris and infected pleural fluid [32]. These fibrinolytic agents include urokinase, streptokinase, and tissue plasminogen activator (tPA). Given that streptokinase has been linked to severe allergic reactions and urokinase is not readily available in the USA, tPA has become the fibrinolytic of choice for many hospitals [6]. Intrapleural fibrinolytic therapies gained popularity after studies concluded that they were superior to traditional chest tube drainage and antibiotics alone [6, 10, 32]. A meta-analysis of 54 studies comparing conservative treatment with antibiotics, drainage, and fibrinolytics found that patients had no in-hospital deaths and a failure rate of <10% [27]. After the establishment of intrathoracic fibrinolytic therapy, studies began comparing patients treated with antibiotics, chest tube drainage and fibrinolytics to those treated with early surgery. The goal of these studies was to determine if the early operative intervention was superior to medical management. Although several retrospective and prospective trials demonstrated a benefit of early VATS [27, 28, 33], randomized controlled trials have reproducibly shown no difference between the two treatment strategies [11, 21, 34, 35]. Specifically, randomized controlled trials by *St. Peter et al.* in 2009, *Sonnappa et al* in 2006, and *Marhuenda et al* in 2014 concluded that chemical debridement with fibrinolytics was equivalent to mechanical debridement with VATS [11, 21, 34]. Endpoints included length of stay, oxygen needs, use of analgesics, complications, and febrile days [36]. Treatment with fibrinolytic therapy was found to be significantly less expensive than treatment with early VATS [21, 34, 35]. The overall failure rate of fibrinolytics was between 10 and 17% [11, 21, 27, 34]. As a result, chemical debridement with fibrinolytics should be offered as a first-line therapy in order to avoid possible morbidity associated with surgery [6, 37, 38]. The APSA treatment algorithm for empyema protocolizes fibrinolytic therapy in order to avoid delays which can decrease the efficacy of these drugs [6, 32, 39]. No clinical trial has demonstrated any lytic drug to be superior. A randomized prospective trial with streptokinase found no advantage of twice-daily dosing over once-daily dosing [40]. Additionally, extended dwell times were found to have no advantage over a one-hour dwell [34]. Finally, deoxyribonuclease (DNase) is a mucolytic agent that, in adults, has been found to improve patient outcomes when combined with lytic agents. Specifically, the Multicenter Intrapleural Sepsis Trail 2 concluded that intrapleural tPA combined with DNase improved fluid drainage, reduced surgical referrals and decreased hospital length of stay when compared to lytic therapy alone [41]. Unfortunately, a 2020 pediatric multi-institutional randomized controlled trial of 97 children demonstrated no benefit from the addition of DNase to intrathoracic tPA. Outcomes for this study included length of stay, chest tube duration, fever duration, ventilatory support needs, surgical referrals and hospital readmissions [42].

b. *Surgical Management*:

    i. *Indications for Surgical Intervention:* Surgical referral is indicated in children where fibrinolytic therapy is not feasible, contraindicated, or unsuccessful. Children who fail fibrinolytic treatment demonstrate persistent fevers, anorexia, hypoxia, and respiratory distress with imaging demonstrating a persistent pleural disease despite low chest tube output. The primary goals of surgery for empyema are to debride the pleural cavity and facilitate lung re-expansion. This is accomplished by draining all the infected fluid and exudate and, when needed, performing a decortication to remove restrictive fibrous tissue or pleural peel around the lung.

    ii. *Thoracoscopic Approach to Empyema:* Minimally invasive VATS is the current gold standard for the surgical management of empyema. Open thoracotomy is associated with higher patient morbidity, postoperative complications and hospital length of stay when compared to VATS [43]. Thoracoscopic debridement and decortication of empyema is performed under general anesthesia with single-lung ventilation (if tolerated). The patient is placed in the lateral decubitus position with the affected side up. The initial trocar is placed at the 4th to 5th intercostal space, anterior axillary line. If tolerated, insufflation can be used to compress the lung and improve working space. An angled scope is inserted and adhesions swept off the chest wall. Under direct visualization, two additional trocars are placed at the 4th to 5th intercostal space, posterior axillary line, and 9th to 10th intercostal space, midaxillary line, in order to establish a triangular position of the port sites. Using suction and irrigation, all the purulent fluid is evacuated, loculations are broken up, and fibrinous exudate debrided. It is important to send samples for culture throughout this procedure in order to help tailor antibiotic choices postop. Next, ringed forceps are used to gently debride the inflammatory peel from the visceral and parietal pleura, with care taken to avoid injuring the lung. Finally, the chest is irrigated and examined for any residual inflammatory tissue or infectious debris. A chest tube is placed at the midaxillary port site and the incisions closed in layers with dissolvable sutures. Postop, the chest tube is examined for an air leak and placed on suction [44].

    iii. *Open Approach to Empyema:* VATS conversion to thoracotomy rate for children with empyema is approximately 1% [27]. For these children and children in which a VATS approach is contraindicated, open thoracotomy can be performed for the management of empyema refractory to fibrinolytics. Once again, the patient is placed in the lateral decubitus position with the affected side up. A posterolateral thoracotomy incision is made, sparing the chest wall muscles if possible. Once access to the chest has been acquired, the steps of the procedure are similar to the thoracoscopic approach.

c. *Postoperative Care and Complications:* The patient can resume a diet immediately postop and should be given a multi-modality pain regimen. The chest tube is maintained until the output is <1 ml/kg/day and there is no air leak. A CXR should be performed on water seal prior to removal of the tube to ensure no pneumothorax or large residual effusion is present and adequate re-expansion of the lung. The complication rate for debridement and decortication of empyema ranges in the literature from 8 to 22% [11, 12, 28]. Possible postoperative complications include the formation of broncho-pleural fistula, pulmonary abscess, pneumothorax, persistent air leak, pneumocele, bronchospasm, need for mechanical ventilation, acute kidney failure, bleeding, sepsis, and pericardial effusion. The postop readmission rate has been reported to be around 7% and is primarily due to recurrent respiratory symptoms or fevers [45].

5. Long-Term Outcomes: Although empyema can cause significant morbidity in children, it is rarely fatal and is associated with good long-term outcomes. Postoperatively at one month, many patients continue to complain of anorexia, intermittent fevers and cough. Additionally, spirometry measurements are abnormal in 35–37% of children and chest X-rays are abnormal in almost all children at one month [45, 46]. Fortunately, studies have demonstrated that, by 1 year, only 2–6% of children have persistent abnormal spirometry, and chest X-rays normalize around 6 months [46] [9]. Neither pulmonary function testing nor time to normalization was found to be different in children receiving VATS compared to those treated with fibrinolytics [13]. Furthermore, at one year, only 4% of parents and 8% of children were noted to have abnormal Pediatric Quality of Life Scores [45].

## Additional Notes

Children with pneumonia can develop parapneumonic effusions when pleural inflammation increases vascular permeability, leading to an influx of inflammatory cells and accumulation of fluid within the thoracic cavity. In contrast, pediatric empyema develops when the fluid collection becomes infected and purulent. The most common underlying etiology of empyema is bacterial transmigration from a concomitant pneumonia or lung infection. Other secondary causes of empyema include chest wall injury or trauma, esophageal injury, bacteremia, infected congenital cyst, neoplasm, subdiaphragmatic infection or postoperative complication following thoracic surgery.

## Study Questions

1. Which of the following stages of empyema development is associated with the development of a "trapped lung"
   a. Exudative stage
   b. Fibrinopurulent stage
   c. Organizing stage

Answer to Question 1: (c.) Organizing stage. In the organizing stage, increased fibroblast production causes thickening of the pleura. An inflammatory rind or peel forms, which can limit the ability of the lung to expand, resulting in a trapped lung.

2. Which of the following is true regarding the three randomized controlled trials comparing fibrinolytic therapy to operative intervention (VATS) for the treatment of empyema?
   a. Length of stay was longer in the VATS groups
   b. The overall failure rates for fibrinolytic therapy is greater than 25%
   c. Treatment with fibrinolytic therapy is significantly less expensive than treatment with early VATS
   d. Fever resolution occurs more quickly in the fibrinolytic treatment group

Answer to Question 2: (c.) Treatment with fibrinolytic therapy is significantly less expensive than treatment with early VATS. Three randomized controlled trials by *St. Peter et al* in 2009, *Sonnappa et al* in 2006, and *Marhuenda et al* in 2014 concluded that chemical debridment with fibrinolytics was equivalent to mechanical debridement with VATS [11, 21, 34]. Endpoints included length of stay, oxygen needs, use of analgesics, complications, and febrile days [36]. Treatment with fibrinolytic therapy was found to be significantly less expensive than treatment with early VATS [21, 34, 35]. The overall failure rate of fibrinolytics was between 10 and 17% [11, 21, 27, 34].

3. Which of the following is an indication of failure of fibrinolytic therapy resulting in need for VATS operative intervention?
   a. Resolution of fevers
   b. High chest tube output
   c. Ongoing oxygen requirements
   d. Lack of improvement on CXR

Answer to Question 3: (c.) Ongoing oxygen requirements. Surgical referral is indicated in children where fibrinolytic therapy is not feasible, contraindicated, or unsuccessful. Children who fail fibrinolytic treatment demonstrate persistent fevers, anorexia, hypoxia, and respiratory distress with imaging, demonstrating a persistent pleural disease despite low chest tube output. Typical CXR finding will lag clinical improvement by several weeks to months. The primary goals of surgery for empyema are to debride the pleural cavity and facilitate lung re-expansion.

## References

1. McIntosh K. Community-acquired pneumonia in children. N Engl J Med. 2002;346(6):429–37.
2. Lee GE, Lorch SA, Sheffler-Collins S, Kronman MP, Shah SS. National hospitalization trends for pediatric pneumonia and associated complications. Pediatrics. 2010;126(2):204–13.
3. Kronman MP, Hersh AL, Feng R, Huang YS, Lee GE, Shah SS. Ambulatory visit rates and antibiotic prescribing for children with pneumonia, 1994-2007. Pediatrics. 2011;127(3):411–8.
4. Shah SS, DiCristina CM, Bell LM, Ten Have T, Metlay JP. Primary early thoracoscopy and reduction in length of hospital stay and additional procedures among children with complicated pneumonia: results of a multicenter retrospective cohort study. Arch Pediatr Adolesc Med. 2008;162(7):675–81.
5. Krenke K, Sadowy E, Podsiadły E, Hryniewicz W, Demkow U, Kulus M. Etiology of parapneumonic effusion and pleural empyema in children. The role of conventional and molecular microbiological tests. Respir Med. 2016;116:28–33.
6. Islam S, Calkins CM, Goldin AB, et al. The diagnosis and management of empyema in children: a comprehensive review from the APSA Outcomes and Clinical Trials Committee. J Pediatr Surg. 2012;47(11):2101–10.
7. Carter E, Waldhausen J, Zhang W, Hoffman L, Redding G. Management of children with empyema: pleural drainage is not always necessary. Pediatr Pulmonol. 2010;45(5):475–80.
8. Grijalva CG, Nuorti JP, Zhu Y, Griffin MR. Increasing incidence of empyema complicating childhood community-acquired pneumonia in the United States. Clin Infect Dis. 2010;50(6):805–13. https://doi.org/10.1086/650573.
9. Calder A, Owens CM. Imaging of parapneumonic pleural effusions and empyema in children. Pediatr Radiol. 2009;39(6):527–37. https://doi.org/10.1007/s00247-008-1133-1.
10. Griffith D, Boal M, Rogers T. Evolution of practice in the management of parapneumonic effusion and empyema in children. J Pediatr Surg. 2018;53(4):644–6.
11. Marhuenda C, Barceló C, Fuentes I, et al. Urokinase versus VATS for treatment of empyema: a randomized multicenter clinical trial. Pediatrics. 2014;134(5):e1301–7.
12. Scarci M, Zahid I, Billé A, Routledge T. Is video-assisted thoracoscopic surgery the best treatment for paediatric pleural empyema? Interact Cardiovasc Thorac Surg. 2011;13(1):70–6.
13. de Benedictis FM, Carloni I, Osimani P, et al. Prospective evaluation of lung function in children with parapneumonic empyema. Pediatr Pulmonol. 2019;54(4):421–7.
14. Segerer FJ, Seeger K, Maier A, et al. Therapy of 645 children with parapneumonic effusion and empyema-A German nationwide surveillance study. Pediatr Pulmonol. 2017;52(4):540–7.
15. Elemraid MA, Thomas MF, Blain AP, et al. Risk factors for the development of pleural empyema in children. Pediatr Pulmonol. 2015;50(7):721–6. https://doi.org/10.1002/ppul.23041.
16. Cagle JG, Zimmerman S, Cohen LW, Porter LS, Hanson LC, Reed D. EMPOWER: an intervention to address barriers to pain management in hospice. J Pain Symptom Manage. 2015;49(1):1–12.
17. Scarci M, Abah U, Solli P, et al. EACTS expert consensus statement for surgical management of pleural empyema. Eur J Cardiothorac Surg. 2015;48(5):642–53.
18. Kurian J, Levin TL, Han BK, Taragin BH, Weinstein S. Comparison of ultrasound and CT in the evaluation of pneumonia complicated by parapneumonic effusion in children. AJR Am J Roentgenol. 2009;193(6):1648–54.
19. Jaffe A, Calder AD, Owens CM, Stanojevic S, Sonnappa S. Role of routine computed tomography in paediatric pleural empyema. Thorax. 2008;63(10):897–902.
20. Sakran W, Ababseh ZD, Miron D, Koren A. Thoracic empyema in children: clinical presentation, microbiology analysis and therapeutic options. J Infect Chemother. 2014;20(4):262–5.

21. St Peter SD, Tsao K, Spilde TL, et al. Thoracoscopic decortication vs tube thoracostomy with fibrinolysis for empyema in children: a prospective, randomized trial. J Pediatr Surg. 2009;44(1):106–11. discussion 111
22. Campbell JD, Nataro JP. Pleural empyema. Pediatr Infect Dis J. 1999;18(8):725–6.
23. Light RW. Parapneumonic effusions and empyema. Proc Am Thorac Soc. 2006;3(1):75–80.
24. Light RW. Parapneumonic effusions and empyema. Clin Chest Med. 1985;6(1):55–62.
25. Gofrit ON, Engelhard D, Abu-Dalu K. Post-pneumonic thoracic empyema in children: a continued surgical challenge. Eur J Pediatr Surg. 1999;9(1):4–7. https://doi.org/10.1055/s-2008-1072203.
26. Karaman I, Erdoğan D, Karaman A, Cakmak O. Comparison of closed-tube thoracostomy and open thoracotomy procedures in the management of thoracic empyema in childhood. Eur J Pediatr Surg. 2004;14(4):250–4.
27. Avansino JR, Goldman B, Sawin RS, Flum DR. Primary operative versus nonoperative therapy for pediatric empyema: a meta-analysis. Pediatrics. 2005;115(6):1652–9.
28. Gates RL, Caniano DA, Hayes JR, Arca MJ. Does VATS provide optimal treatment of empyema in children? A systematic review. J Pediatr Surg. 2004;39(3):381–6.
29. Balfour-Lynn IM, Abrahamson E, Cohen G, et al. BTS guidelines for the management of pleural infection in children. Thorax. 2005;60(Suppl 1):i1–21. https://doi.org/10.1136/thx.2004.030676.
30. Shen KR, Bribriesco A, Crabtree T, et al. The American Association for Thoracic Surgery consensus guidelines for the management of empyema. J Thorac Cardiovasc Surg. 2017;153(6):e129–46.
31. Lin CH, Lin WC, Chang JS. Comparison of pigtail catheter with chest tube for drainage of parapneumonic effusion in children. Pediatr Neonatol. 2011;52(6):337–41.
32. Hanson SJ, Havens PL, Simpson PM, Nugent ML, Wells RG. Intrapleural alteplase decreases parapneumonic effusion volume in children more than saline irrigation. Pediatr Pulmonol. 2015;50(12):1328–35.
33. Grewal H, Jackson RJ, Wagner CW, Smith SD. Early video-assisted thoracic surgery in the management of empyema. Pediatrics. 1999;103(5):e63. https://doi.org/10.1542/peds.103.5.e63.
34. Sonnappa S, Cohen G, Owens CM, et al. Comparison of urokinase and video-assisted thoracoscopic surgery for treatment of childhood empyema. Am J Respir Crit Care Med. 2006;174(2):221–7.
35. Kurt BA, Winterhalter KM, Connors RH, Betz BW, Winters JW. Therapy of parapneumonic effusions in children: video-assisted thoracoscopic surgery versus conventional thoracostomy drainage. Pediatrics. 2006;118(3):e547–53.
36. Krenke K, Peradzyńska J, Lange J, Ruszczyński M, Kulus M, Szajewska H. Local treatment of empyema in children: a systematic review of randomized controlled trials. Acta Paediatr. 2010;99(10):1449–53.
37. Cameron R, Davies HR. Intra-pleural fibrinolytic therapy versus conservative management in the treatment of adult parapneumonic effusions and empyema. Cochrane Database Syst Rev. 2008;(2):CD002312.
38. Redden MD, Chin TY, van Driel ML. Surgical versus non-surgical management for pleural empyema. Cochrane Database Syst Rev 2017;3:CD010651.
39. Gasior AC, Knott EM, Sharp SW, Ostlie DJ, Holcomb GW, St Peter SD. Experience with an evidence-based protocol using fibrinolysis as first line treatment for empyema in children. J Pediatr Surg. 2013;48(6):1312–5.
40. Wang JN, Yao CT, Yeh CN, et al. Once-daily vs. twice-daily intrapleural urokinase treatment of complicated parapneumonic effusion in paediatric patients: a randomised, prospective study. Int J Clin Pract. 2006;60(10):1225–30.
41. Rahman NM, Maskell NA, West A, et al. Intrapleural use of tissue plasminogen activator and DNase in pleural infection. N Engl J Med. 2011;365(6):518–26.

42. Livingston MH, Mahant S, Connolly B, et al. Effectiveness of intrapleural tissue plasminogen activator and dornase alfa vs tissue plasminogen activator alone in children with pleural empyema: a randomized clinical trial. JAMA Pediatr. 2020;174(4):332–40.
43. Jaffé A, Cohen G. Thoracic empyema. Arch Dis Child. 2003;88(10):839–41.
44. Atlas of Pediatric Laparoscopy and Thoracoscopy. 2nd ed. Elsevier; 2021.
45. Cohen E, Mahant S, Dell SD, et al. The long-term outcomes of pediatric pleural empyema: a prospective study. Arch Pediatr Adolesc Med. 2012;166(11):999–1004.
46. Maffey A, Colom A, Venialgo C, et al. Clinical, functional, and radiological outcome in children with pleural empyema. Pediatr Pulmonol. 2019;54(5):525–30.

# Vascular Rings and Aberrant Brachiocephalic Trunk

**Gerhard Ziemer and Cherish Boehm**

**Key Points**

1) Vascular rings can develop in many different individual patterns because they result from abnormal persistence of embryologic structures from the multiple parts of the right and left aorta dorsal and ventral segments, as well as the interconnecting pharyngeal arches and arteries.
2) Not all vascular rings are symptomatic, but when symptoms are present, they are due to compression of the trachea or the esophagus by the abnormal structures, resulting in dyspnea or dysphagia.
3) In general, the best operative approach for any vascular ring is a lateral thoracotomy performed on the side of the patient that has a descending aorta. Different approach is possible.
4) Advanced imaging techniques result in more vascular rings being diagnosed, often during the prenatal period, and with excellent localization of anatomy, leading to discussion for treatment without symptoms.

## 1) Introduction

At first sight vascular anomalies causing compression of neighboring structures do not appear to be simply categorized because of their diversity.

The development of the aortic arch and its branches from two dorsal and two ventral parts and their interconnecting pharyngeal arches/arteries is substantiated by embryology leading to numerous possibilities of ring development, but they have

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only isolated clinical relevance (Fig. 1). A definite simplified classification serving >95% of the clinically described variations is as follows:

A) Double aortic arch (Fig. 2)
B) Right-sided aortic arch/left-sided arterial ligament
C) Aberrant origin of the right-sided brachiocephalic trunk causing tracheal stenosis.

The common feature between these anomalies is the compression of the neighboring structures such as the trachea and esophagus causing shortness of breath and sometimes difficulties in swallowing. In infancy with predominantly liquid food as mother's milk/formula and a rather soft trachea, the most common symptoms are respiratory. With increasing age, difficulties in swallowing/dysphagia increase with the introduction of solid food causing more pronounced dysphagia. However,

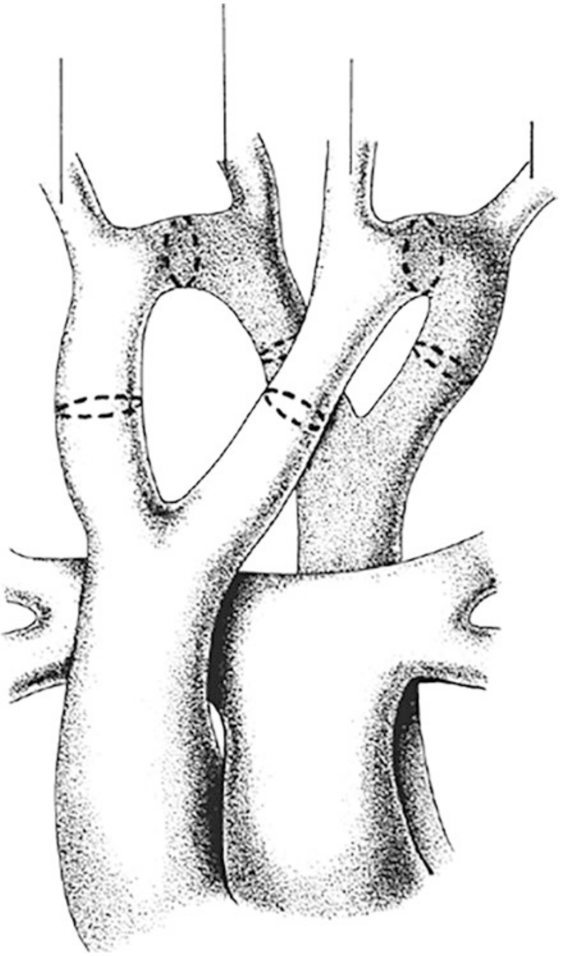

**Fig. 1** Primitive aortic arch, the dashed lines indicate all possible interruptions, which explain all possible forms of normal and abnormal aortic arch and vascular rings

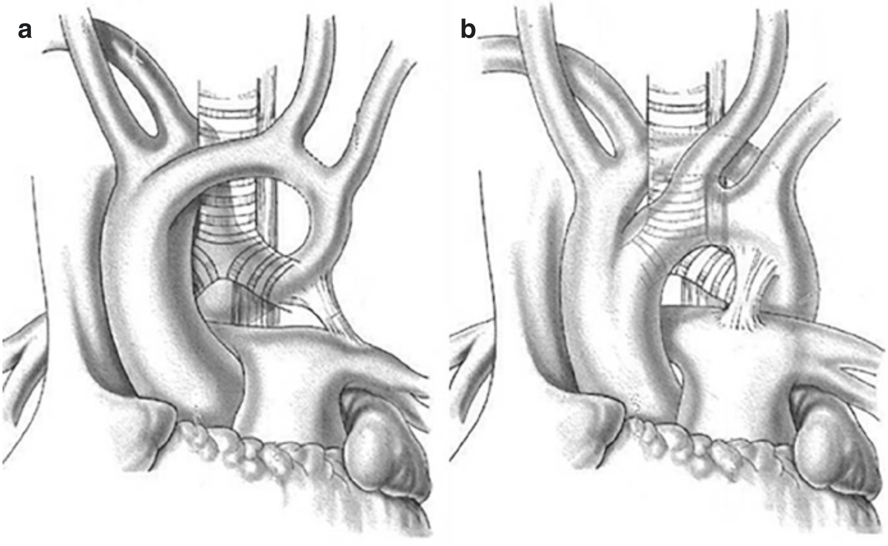

**Fig. 2** Double aortic arch. (**a**) The most common form of double aortic arch: right-sided aortic arch with a hypoplastic left arch. The hypoplastic arch can become extremely hypoplastic or even atretic either between the supra-aortic vessels or distal to them (**b**). The rare form of a dominant left-sided aortic arch with a hypoplastic retroesophageal segment of the right-sided arch with the descending aorta on the left

patients who never experienced normal swallowing before, may not be able to report their objective dysphagia.

Modern imaging tools, such as computed tomography (CT)/magnetic resonance imaging (MRI), improve the accuracy of localizing defects. The timely and up-to-date implementation of surgery brings about immediate regression and abolishment of symptoms in most patients with excellent long-term results.

The variations in the course of the thoracic aorta do not necessarily cause symptoms. Contrary to other congenital heart diseases, anomalies of intrathoracic great vessels are only operated upon when clinical symptoms are present. However, there are two exceptions:

A. An asymptomatic vascular anomaly associated with intracardiac defects requiring surgery
B. An asymptomatic complete double aortic arch as it will become symptomatic with age and growth, but surgery is much easier in neonates and infants.

A new clinical challenge is the increasing number of asymptomatic vascular rings being diagnosed during fetal life.

The radical resection of the encircling vascular and fibrotic structures is recommended. The importance of the Kommerell diverticulum as an independent cause for recurrent symptoms has been recognized over the years and addressed now for primary surgery.

Video-assisted thoracoscopic repair as well as totally endoscopic robotic-assisted approach is reported. These approaches, however, do not meet the abovementioned principles of radical resection.

Symptomatic pulmonary artery sling is not part of this chapter, as it most often requires additional tracheal surgery, which nowadays is performed via sternotomy, employing extracorporeal circulation, and therefore done in pediatric cardiac surgery units.

### 2) Aim of Surgery for Vascular Rings

In our view, with the exception of an aberrant brachiocephalic trunk, for all compressing thoracic vascular anomalies, the approach, a lateral thoracotomy, should always be performed on the side of the descending aorta. The argument in favor of such an approach is to obtain safe intraoperative control of the structures to be cut, especially of the descending aorta. A complete resection starts at the descending aorta and extends beyond the esophagus on the other side if not limited by specific anatomic structures/details we are not aware of before operation.

The resection of a patent vascular segment can be carried out by side bite clamping or under certain circumstances in cross-clamping of the descending aorta above and below the structure to be resected. To perform a continuous running suture to close the resected structure, flush to the aortic wall should take only a few minutes, and for that there is no need for further protective measures like including a left heart assist.

It must be mentioned that most surgeons perform a division of any vascular ring irrespective of the side of the descending aorta via a left-sided posterior thoracotomy, which occasionally leads to safety issues when following the principles of radical resection. An advantage may be the option of re-implantation of an aberrant subclavian artery on the side of thoracotomy.

The immediate intraoperative sign of effectively having taken care of the symptomatic vascular ring is the significant retraction of the divided ring structures.

Vascular rings are associated only rarely with a primary structural tracheal stenosis caused by hypoplasia in contrast to a pulmonary (artery) sling. A certain degree of tracheomalacia can be the reason for a residual stridor in infants in the first postoperative month and in most cases does not require additional surgery.

### 3) Double Aortic Arch/Complete Vascular Ring

For all types of vascular rings, the following anatomic and hemodynamic factors have to be taken into consideration when planning for surgery:

A) Position of the descending aorta, especially in relation to the esophagus
B) Position of the dominant aortic arch
C) Possibility of transection of currently patent vascular structures.

The sidedness of the descending aorta in relation to the esophagus is important because the access to the aorta can be challenging when the esophagus has to be passed.

The proportions of size between the anterior and posterior parts of the arches vary. Double aortic arch may consist of two equally and well-developed arches. Commonly there is a dominant right aortic arch with a hypoplastic left arch.

In case of a double aortic arch, the nondominant part should be divided. If there is no side preference reported, accessibility dictates the arch to be transected.

Surgery of a balanced (co-dominant) double aortic arch follows the principles of careful retroesophageal dissection and resection as mentioned above. Prior to actual division, the strength of the right and left carotid and radial pulses should be evaluated with the vascular clamps applied. In the absence of any changes in proximal and distal blood pressure and pulse curves, even the bigger of the two aortic arches can be divided. To divide the bigger of two arches in double aortic arch may be thought of, if the bigger arch is the one with a retroesophageal course. Precise documentation is required.

In case of a dominant aortic arch with retroesophageal course, a simple transection of the anterior hypoplastic ring segment may not be sufficient to relieve all, but it may diminish some clinical symptoms.

### 4) Incomplete Vascular Ring

Right-sided aortic arch with left-sided Ductus Botalli/Ligamentum Arteriosum

The right-sided aortic arch is curving to the right of the trachea. There are three theoretical possible varieties, two of which may create a vascular ring. The fundamental structure is always the retroesophageal vascular structure or its remnant (arterial duct vs. arterial ligament).

The first variety, the most commonly seen patologic form (65%), is the combination of an aberrant left subclavian artery and an arterial duct located between the left pulmonary artery and aortic isthmus. A short aortic segment is addressed here indeed as the isthmus from which the left subclavian artery and the duct arise (Fig. 3).

Both structures can arise separately from the aorta or jointly from a Kommerell diverticulum. The existence of a Kommerell diverticulum is not obligatory for this kind of symptomatic vascular ring.

The second variety (33%) is the mirror image of the normal left aortic arch: Right aortic arch with a left-sided brachiocephalic trunk giving rise to the left subclavian artery which runs regularly to the left and anteriorly of the trachea. The ring is created by a retroesophageal duct respectively ligamentum arteriosum, only, coming off the right sided isthmus.

The third variety, also with a mirror image branching, does not create a vascular ring, as the ductus originates from the left subclavian artery directly. This form occurs sporadically (2%) and is associated with a tetralogy of Fallot without causing any vascular ring type symptoms.

### 5) Kommerell Diverticulum

The Kommerell diverticulum is named after Burckhard Kommerell, staff radiologist at Charité Hospital, Berlin, Germany, in the 1930s. It is a remnant of the dorsal right or left root of the aortic arch as a pendant to the contralateral existing complete aortic arch (Fig. 3). This structure, like when first described, is not referring to a certain kind of vascular ring but can occur, not obligatory, in a number of anomalies. The diverticulum is an aorta-sized outpouching of the descending aorta

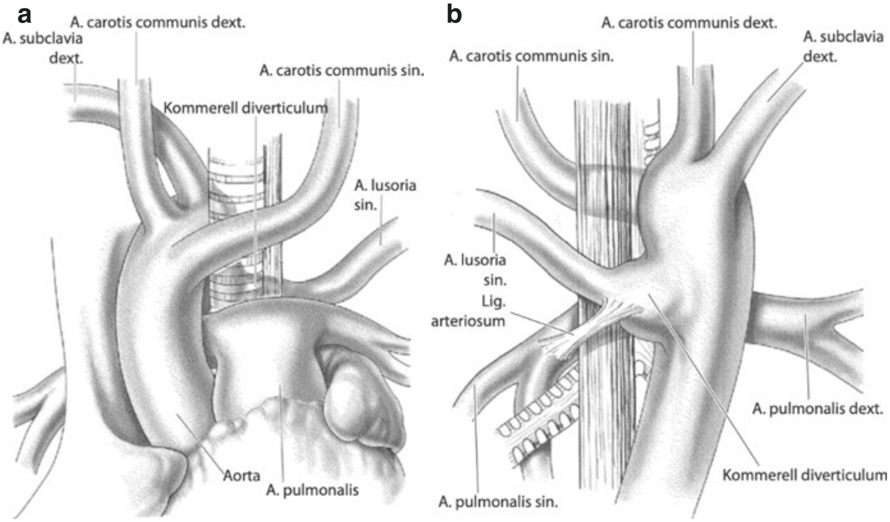
a
A. subclavia dext.
A. carotis communis dext.
A. carotis communis sin.
Kommerell diverticulum
A. lusoria sin.
Aorta
A. pulmonalis
b
A. carotis communis sin.
A. carotis communis dext.
A. subclavia dext.
A. lusoria sin.
Lig. arteriosum
A. pulmonalis dext.
Kommerell diverticulum
A. pulmonalis sin.

**Fig. 3** Incomplete vascular ring: View of a Kommerell diverticulum with a right-sided aortic arch and aberrant left subclavian artery. View from ventral (**a**) and view from dorsal (**b**)

ending anteriorly in a nondominant/atretic aortic arch or in an arterial duct and aberrant subclavian artery (lusorian artery). It was originally described in a patient with a left-sided aortic arch and retroesophageally located right subclavian artery.

The diverticulum can cause obstruction as part of a vascular ring compression, or probably only because of its retroesophageal volume. This is confirmed by the fact that a Kommerell diverticulum is a frequent cause of a symptomatic recurrence or persistence of symptoms following surgery of division of a vascular ring without resection of the diverticulum.

6) Less Common Findings of a Vascular Ring

The diversity of these findings published mostly as a pathological case report or as an isolated clinical case is quite understandable given the complex embryologic development (Fig. 1). As these forms account for only about 1% of the clinical cases with vascular ring malformation, they cannot be focused on individually.

7) Compression Caused by an Aberrant Origin of the Brachiocephalic Trunk/Innominate Artery

This anomaly is caused by an atypical distal take-off of the innominate artery. It does not form a vascular ring. The artery leaves the arch distally posteriorly. Coming from the left, it takes a course ventral to the right around the trachea heading into a posterior direction. This at times causes a compression of the trachea. The ventral indentation of the trachea seen by bronchoscopy is pathognomonic for this anomaly; no other vascular anomaly causes an isolated indent like that (Fig. 4).

For minor stridor symptoms, the treatment is mostly conservative because with the development of the cartilaginous rings during the second and third year of life symptoms diminish or may even disappear completely. This may be also due to the changing size relation of the structures with growth. A symptomatic, significant tracheal stenosis (70–80%) or apneic spells during infancy and early childhood should be an indication for surgical treatment.

This anomaly is mainly seen in infants and causes a typical stridor. Even episodes of short apnea are described.

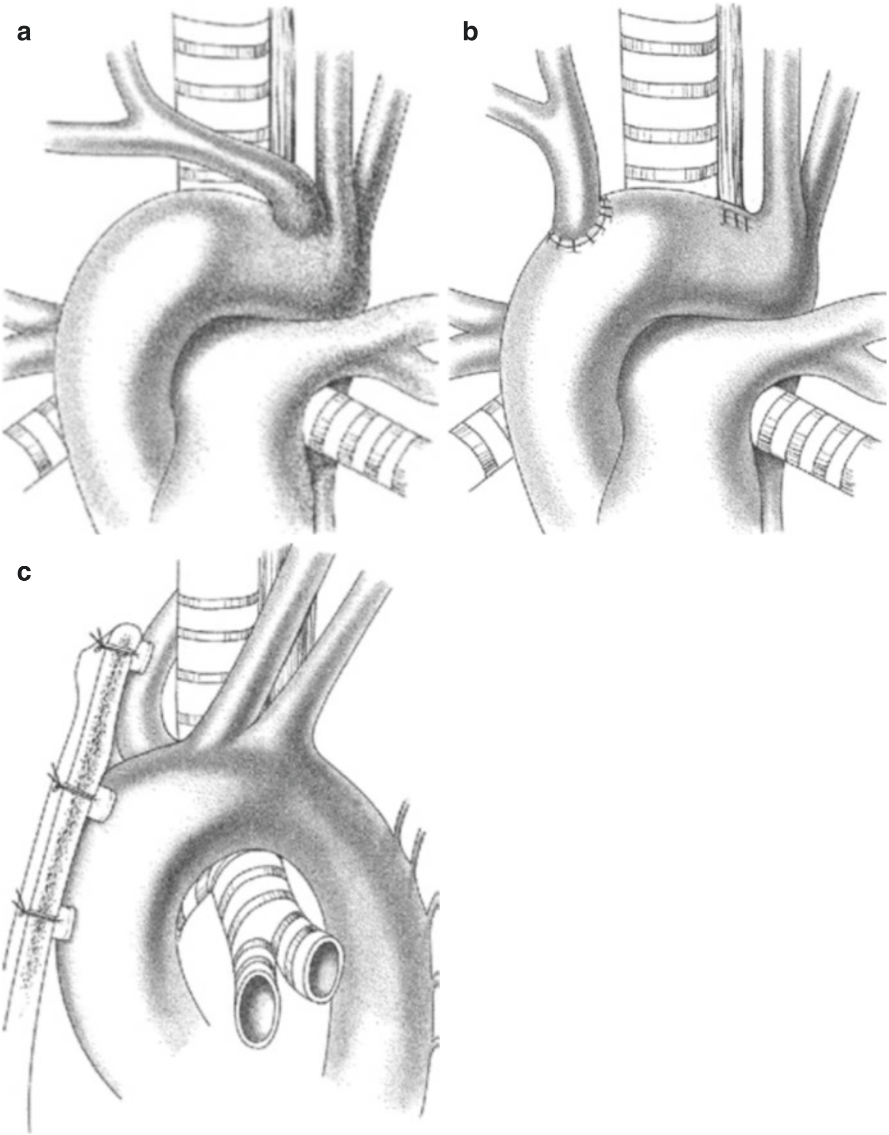

**Fig. 4** Aberrant brachiocephalic trunk/innominate artery. (**a**, **b**) Anterior translocation. (**c**) Anterior aortotruncopexy

An established therapy is the classical ventral truncopexy—tacking the innominate artery to the sternum (Fig. 4c)—or, as a more complex alternative, the transsternal resection followed by a proximal re-implantation of the innominate artery into the proximal aortic arch or the distal ascending aorta (Fig. 4b), our preferred choice. We always employ extracorporeal circulation and mild hypothermia for this procedure. During the procedure, Near Infrared Spectroscopy (NIRS) is used to monitor brain perfusion.

For truncopexy, the original surgical approach described by Gross is the left thoracotomy. Other authors prefer a right anterolateral thoracotomy with a inframammary incision line. When we do a pexy rather than a translocation, we use a median partial upper sternotomy. The argument in favor of such access is the safe anchoring of the transsternal sutures.

We use three U-figured pledged sutures (3.0 or 4.0). Instead of a mere truncopexy, we perform an aortotruncopexy (Fig. 4c): we place the first suture subadventially at the distal ascending aorta, the second at the base of the innominate artery, and the third at the distal trunk just above the innominate vein. The sutures are fed transsternally through the right part of the sternum and tightened fast after the removal of the retractor. With that the pulse oximeter measurement applied at the right arm should be unchanged, and the bronchoscopy, which should be performed obligatorily, should show an expansion of the tracheal lumen, ideally with disappearance of the pulsations. Late re-sternotomies for any reason may find the aortotruncopexy loosened without recurrence of tracheal symptoms.

## Additional Notes

Further notes on the Kommerell diverticulum: An increasing number of case reports describe an aneurysmatic dilatation of an originally asymptomatic diverticulum occasionally leading to aortic dissection later in life. For this reason, also, it is highly recommended to resect a diverticulum during primary surgery of any vascular ring if present. Endovascular procedures may be able to exclude a retroesophageal aneurysm from the circulation, but they may not relieve the gastrointestinal symptoms if present before.

## Study Questions

1) What is a common stage in life for a child with a vascular ring to begin experiencing symptoms of dysphagia?

   a. Shortly after birth
   b. With transition from liquid to solid foods

   c. During the growth spurt of adolescence
   d. With scarring of structure following surgical correction

Answer to Question 1: (b.) With transition from liquid to solid foods. Clinically significant vascular rings result in compression of neighboring structures such as the trachea and esophagus. This causes the typical symptoms of shortness of breath and dysphagia. During infancy, when babies consume liquid food and the trachea is relatively soft, the most common symptom is shortness of breath. With increasing age, the introduction of solid food can result in more pronounced dysphagia. Growth of the trachea with stronger cartilaginous rings and greater diameter can improve respiratory symptoms.

2) Which lesion can cause obstructive symptoms after surgical correction of a vascular ring?

   a. Duplicate aortic arch
   b. Aneurysmal dilatation of a Kommerell diverticulum
   c. Ligamentum arteriosum
   d. Aberrant brachiocephalic trunk/innominate artery

Answer to Question 2: (c.) Aneurysmal dilatation of a Kommerell diverticulum. It is an aorta-sized outpouching of the descending aorta ending anteriorly in a non-dominant/atretic aortic arch or in an arterial duct and aberrant subclavian artery (lusorian artery). The Kommerell diverticulum is a frequent cause of a symptomatic recurrence or persistence of symptoms following surgery of division of a vascular ring without resection of the diverticulum.

## Further Reading

Ziemer G, Kaulitz R. Congenital anomalies of the great vessels. 21.2.2 Aortic anomalies causing compression of neighboring structures. In: Cardiac surgery. Operations on the heart and great vessels in adults and children, Ziemer G, Haverich A, editor. 1st engl edition. 1158 pp. Berlin: Springer; 2017, p. 688–99.

# Part VII
# Common Adolescent Problems

# Gynecomastia

**Ashley Jordan, Joseph DeSantis, and Christian Kauffman**

**Key Points**

1) Gynecomastia is the enlargement of breast tissue in males.
2) Most common cause is idiopathic, but thorough workup may be required to rule out pathologic etiology.
3) Surgical treatment includes a variety of options from liposuction to direct excision of breast tissue and skin.

Gynecomastia is the pathologic enlargement of the breast in males. It can occur in neonates, early adolescence, and late adulthood. Most cases are idiopathic however, it also has physiologic, pathologic, and pharmacological causes. It is important to differentiate gynecomastia from pseudogynecomastia which is fatty hypertrophy of the breasts. A thorough history and physical exam are essential components in the workup of gynecomastia. Patients often present with a firm or rubbery mass extending concentrically and symmetrically from the nipple. Masses are typically bilateral but can be unilateral. Laboratory and radiographic studies are usually unnecessary although in some cases prove to be helpful. Initial treatment is observation as many cases of gynecomastia in pubescent males resolve without intervention. If surgery is deemed necessary, the surgical procedure is determined by the amount of fibrinous breast tissue versus adipose tissue, breast ptosis, and the amount of redundant skin. Operative strategies are best managed by a board-certified plastic surgeon and include liposuction, peri-areolar excision, circumareolar skin excision, transverse

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skin resection with nipple-areolar preservation on a dermal pedicle, transverse skin resection with free nipple/areola graft and a keyhole skin excision. The most common complications include hematoma and insufficient resection.

1. Epidemiology
    a. Incidence:
        i. Typically occurs at three time periods: neonatal, early adolescence, and late adulthood.
        ii. Overall incidence 32–36%.
    b. Etiology—Multifactorial
        i. Idiopathic—most common cause.
        ii. Physiologic
            1. Neonatal—secondary to circulating maternal estrogens.
            2. Pubertal—imbalance of estradiol and testosterone.
                a. Up to 65% of adolescent boys develop gynecomastia. Often self-limited with resolution in 6–18 months.
            3. Elderly (>65 years of age)—decline in testosterone, peripheral aromatization of testosterone to estrogen, shift in ratio of testosterone to estrogen.
        iii. Obesity—pseudogynecomastia (fatty hypertrophy).
        iv. Pathologic
            1. Renal failure, hyperthyroidism, hypothyroidism, liver disease, adrenal tumors, testicular tumors (increased estrogen), congenital or acquired hypogonadism (decreased testosterone—Klinefelter's syndrome, orchiectomy).
        v. Pharmacologic
            1. Alcohol, amphetamines, chemotherapeutic agents, cimetidine, digitalis, haldol, hydroxyzine, isoniazid, methyldopa, marijuana, opiates, phenothiazines, progestins, reserpine, spironolactone, tricyclic antidepressants, anabolic steroids.
2. Pathophysiology:
    a. Imbalance in estradiol and testosterone causes glandular proliferation, ductal epithelial hyperplasia, ductal elongation and branching, and proliferation of periductal fibroblasts.
    b. American Society of Plastic Surgeons (ASPS) Sanctioned Classification Scheme:
        i. Grade I: Small breast enlargement with a localized button of tissue around the areola.

      - ii. Grade II: Moderate breast enlargement exceeding areolar boundaries with edges that are indistinct from the chest.
      - iii. Grade III: Moderate breast enlargement exceeding areolar boundaries with edges that are distinct from the chest with skin redundancy.
      - iv. Grade IV: Marked breast enlargement with skin redundancy and feminization of the breast.

   - c. Histologic Classification
      - i. Florid—increase in ductal tissue and vascularity. Duration less than 4 months.
      - ii. Intermediate—mixture of florid and fibrous. Seen between 4 and 12 months.
      - iii. Fibrous—stromal fibrosis, few ducts. Usually present when gynecomastia lasts more than 1 year.

   - d. Cancer risk
      - i. Klinefelter's syndrome: risk increases 60-fold.
      - ii. No increased risk of cancer for patients who do not have Klinefelter's syndrome.

3. Clinical features
   - a. Gynecomastia: Patients present with a firm or rubbery mass extending concentrically and symmetrically from the nipple.
      - i. Typically bilateral (75%) but can be unilateral.
      - ii. Patients often experience extreme psychosocial and self-esteem issues.
      - iii. Patients may be bullied and teased by their peers, avoid changing in locker rooms or in front of others, avoid swimming pools, the beach and other activities involving the possibility of exposing their chest.
   - b. Pseudogynecomastia: little to no resistance when the area is pinched, feels like diffuse fat instead of a discrete mass.

4. Diagnosis: A detailed history and physical examination are the most important aspects in the workup for gynecomastia.
   - a. History should include:
      - i. Age of onset and duration of gynecomastia.
      - ii. Pain and/or tenderness in the breasts.
      - iii. Changes in the size of the breast and the rate of change.
      - iv. Nipple symptoms: tenderness, pain, discharge.
      - v. Drug use: current use and past use; medically prescribed and recreational drugs.
      - vi. Other medical problems.
      - vii. Family history of gynecomastia.
      - viii. Review of symptoms:

1. Organs system changes associated with gynecomastia include:
    a. Thyroid: weight loss, tremor, anxiety.
    b. Genital/Testicular: Hypogonadism, impotence, decreased libido, change in the size of the testes.
    c. Liver, renal, adrenal, pulmonary, pituitary, and prostate can also be affected.

b. Physical Examination:
   i. Breast: Perform the “pinch test” to determine the nature and consistency of the breast tissue (glandular, fibrous, solid, fatty), note the laterality, asymmetry, volume, area, ptosis, areola size, presence or absence of isolated masses, tenderness.
   ii. Chest: Evaluate for chest wall deformities.
   iii. Axillae: Evaluate for the presence of palpable lymph nodes.
   iv. Abdomen: Palpate for adrenal and/or liver enlargement.
   v. Thyroid: Assess for enlargement.
   vi. Genital/Testes: Determine Tanner staging, examine for asymmetry, masses, cryptorchidism, enlargement, or atrophy.

c. Laboratory evaluation
   i. Healthy pubertal children with a normal physical exam (other than physiologic gynecomastia) typically do not require further workup.
   ii. Patients with feminizing characteristics (small testis, sparse or absent male pubic hair distribution) should undergo endocrine evaluation.
      1. If a marfanoid body habitus is associated with the feminization, Klinefelter syndrome must be ruled out and the patient should be referred to an endocrinologist.
   iii. Prepubertal, but not neonatal, gynecomastia requires additional workup.
   iv. Tests to consider: human chorionic gonadotropin (hCG), luteinizing hormone (LH), testosterone, estradiol, Free T4, thyroid stimulating hormone (TSH), prolactin, growth hormone (GH).

d. Radiographic studies: consider ordering an ultrasound if there is the presence of prepubertal gynecomastia or a suspicious breast, axillary or testicular mass or significant asymmetry.

e. Consultations to consider:
   i. Plastic surgeon, endocrinologist, urologist, psychiatrist.

5. Treatment
   a. Initial Measures—gynecomastia is not uncommon in adolescent males and the majority are idiopathic.

    - i. Observation is the first step in the management of new gynecomastia in a pubescent male.
    - ii. Gynecomastia in prepubescent patients or abnormal findings in the history and physical may warrant additional workup.

- b. Subsequent Measures
    - i. Medications or recreational drugs that are known to cause gynecomastia should be discontinued.
    - ii. Testosterone can be considered in cases of hypogonadism.
    - iii. Estrogen blockers such as tamoxifen and raloxifene are typically not used in teenagers and are not approved in the USA for the treatment of gynecomastia.

- c. Preoperative Preparation
    - i. Patient age and stability of gynecomastia must be weighed against the physical and psychological burden.
    - ii. Younger age and enlarging breast tissue increase the risk of recurrence.
    - iii. The surgical procedure is determined by the amount of breast/fibrous tissue (true gynecomastia) versus adipose tissue (pseudogynecomastia), breast ptosis, and the amount of redundant skin.
    - iv. Patients should be evaluated by a plastic surgeon to determine the best operative treatment.

- d. Operative Treatment
    - i. Liposuction
        1. Consider liposuction alone in a patient presenting with gynecomastia consisting primarily of adipose tissue with minimal redundant skin and no ptosis.
        2. Scars are minimized.
        3. The use of ultrasonic liposuction, as an adjunct to suction-assisted liposuction, may be helpful in improving adipose removal.
        4. Liposuction will not remove breast/fibrous tissue and will not reduce redundant skin.
        5. Breast ptosis may worsen after liposuction.
    - ii. Peri-areolar excision
        1. Incision is relatively hidden at the lower interface between the areola and chest skin allowing breast/fibrous tissue to be directly removed.
        2. Contouring of the surrounding adipose tissue with liposuction is beneficial in some patients.
        3. Does not address redundant skin or breast ptosis.
        4. The most common procedure for gynecomastia done in teenagers.

iii. Circumareolar skin excision

    1. Similar to peri-areolar approach.
    2. Excision of skin in a donut pattern which places the scar around the areola.
    3. Allows removal of a small amount of redundant skin and direct resection of breast/fibrous tissue.
    4. Unlike liposuction and peri-areolar excision, an areolar reduction can also be performed with this technique.
    5. This technique does not address ptosis and skin removal is limited. Widened or hypertrophic scars can occur.

iv. Transverse skin resection with nipple-areolar preservation on a dermal pedicle.

    1. Removes breast/fibrous tissue, redundant skin and corrects breast ptosis. The areola width can also be reduced.
    2. A scar is created most often at the inframammary crease.
    3. Ischemia and necrosis of the nipple-areolar complex may occur but is uncommon.

v. Transverse skin resection with free nipple/areola graft

    1. Best for patients with redundant skin and severe ptosis in which a dermal pedicle would be too long to support the blood supply to the nipple-areolar complex or the pedicle is too bulky to create a flat masculine chest.
    2. Free nipple grafts can have delayed healing. There is a risk of complete loss of the free nipple graft although uncommon.

vi. Keyhole skin excision

    1. Used when there is a vertical and horizontal excess of skin.
    2. Used with either a dermal pedicle or free nipple graft.
    3. The design can create a “pointed” look and is not commonly performed.

e. Postoperative Treatment

i. A compression garment, typically a vest, is worn for 4–6 weeks after surgery.
ii. Physical restriction, especially of the upper body, is mandated for 4–6 weeks.
iii. Suction drains are typically used except when only liposuction is performed, or the resection is very limited.
iv. The final appearance of the chest may not be apparent for up to a year, so revisions are typically postponed until at least 6 months after surgery.

- f. Complications
  - i. Hematoma/seroma
    1. Most common early complication.
    2. Incidence of 5–10%.
    3. Drainage is essential to prevent scarring which results in poor contour and asymmetry.
  - ii. Under resection of tissue
    1. Most common late complication.
    2. Typically managed as a minor procedure to resect more tissue under local anesthesia.

## Additional Notes

The evaluation of the adolescent gynecomastia patient is complex with no firm rules regarding surgical timing. Gynecomastia most often resolves without operative intervention. The varied surgeries for gynecomastia indicate that its treatment is not a one operation fits all. The surgical techniques reviewed are best managed by a plastic surgeon.

## Questions

1. Which of these patients most likely represents idiopathic gynecomastia?
   - A. A 70-year-old male with new onset unilateral breast enlargement
   - B. A 30-year-old body builder with breast development
   - C. A 13-year-old male with bilateral painful breast masses
   - D. A 45-year-old-male with ptotic breasts following gastric bypass

The answer is C. An adolescent male with painful breast enlargement is typical of idiopathic gynecomastia.

2. A 10-year-old male presents with new onset breast development. Which of the following is *not* an appropriate next step in treatment?
   - A. Testicular exam
   - B. Ultrasound of breasts
   - C. Excisional biopsy
   - D. Testosterone level

The answer is C. Workup may include a testicular exam, ultrasound, and testosterone level. Surgery in a 10-year-old including biopsy is not indicated until thorough work-up has been completed.

3. A 20-year-old male presents with gynecomastia following a 140 lb weight loss after gastric bypass. Severe nipple ptosis and redundant chest skin are noted on physical exam. What is the best surgical approach?

   A. Ultrasound assisted liposuction
   B. Circumareolar skin excision and resection of fibrous tissue
   C. Keyhole resection
   D. Transverse skin excision with free nipple graft

The answer is D. The most appropriate surgical approach in a gynecomastia patient with severe ptosis and redundant tissue must involve skin resection. The best method is a transverse excision and free nipple graft.

## Further Reading

Hammond DC. Surgical correction of gynecomastia. Plast Reconstruct Surg. 2009;124(1):61e–8e.
Janis JE, editor. Essentials of plastic surgery. CRC Press; 2019.
Jones GE. Bostwick's plastic and reconstructive breast surgery. Thieme; 2020.
Kasielska A, Antoszewski B. Effect of operative treatment on psychosocial problems of men with gynaecomastia. Polski przeglad chirurgiczny. 2011;83(11):614–21.
Kinsella C Jr, Landfair A, Rottgers SA, Cray JJ, Weidman C, Deleyiannis FW, Grunwaldt L, Losee JE. The psychological burden of idiopathic adolescent gynecomastia. Plast Reconstruct Surg. 2012;129(1):1–7.
Mast BA, editor. Plastic surgery: a practical guide to operative care. Thieme; 2019.
Thaller SR. Grabb and Smith's plastic surgery. Plast Reconstruct Surg. 2014;133(5):1311–2.

# Pectus Deformities

## Daniel P. Croitoru

**Key Points**

1) Chest wall deformities comprise a spectrum of deformities from rare life-threatening deformities to those that have minimal symptoms.
2) Pectus excavatum is the most common deformity and can have associated symptoms and rarely requires urgent repair.
3) Pectus excavatum when symptomatic is most commonly repaired by the minimally invasive pectus excavatum repair (Nuss technique) or by open repair (Ravitch technique).
4) Pectus carinatum is the second most common deformity and may be corrected without operation.

Chest wall deformities manifest as a spectrum of congenital abnormalities of alterations in the position and shape of the anterior chest wall including pectus excavatum and pectus carinatum, the most common abnormalities, to the extremely rare abnormalities of aplasia and dysplasia of the chest wall which include thoracic ectopic cordis, sternal cleft, Poland syndrome, Jeune asphyxiating dystrophy, and Jarcho–Levin spondylocostal dysplasia syndrome. Acquired abnormalities may also occur following thoracic surgery procedures in early infancy including diaphragmatic hernia repair.

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## Classification of Chest Wall Deformities

1. Pectus excavatum: “funnel chest”—sunken/concave depression of the sternum and costochondral cartilages and is the most common chest wall deformity (90+%).
2. Pectus carinatum: “pigeon chest”—outward protrusion of the sternum and costochondral cartilages ribs with a bird-like appearance of the chest.
3. Combined deformity: asymmetric or mixed deformity with tilt of the sternum off of the coronal plane, flaring of the ribs, and disordered alignment of the ribs.
4. Chest wall aplasia and dysplasia (rare in incidence): missing or malformed components of the rib cage including sternum, vertebrae, hemivertebrae, absent ribs and cartilage, fused ribs, absent clavicles, and can also include aplasia of the soft tissue of the chest.
    a. Sternal cleft—midline non-fusion of sternal elements.
    b. Poland syndrome—unilateral chest wall deficit of costochondral cartilages, ribs and can have dysplasia or aplasia of pectoral muscles, breast, and ipsilateral limb.
    c. Jeune’s asphyxiating thoracic dystrophy is characterized by failure of rib growth and a bell-shaped chest with a narrow apex. Patients often have difficulty handling secretions and require tracheostomy.
    d. Jarcho–Levin Syndrome (spondylocostal dysplasia) is characterized by shortened deformed thoracic spine and dysplastic ribs and has a high mortality.
    e. Pentalogy of Cantrell also has a very high mortality and includes:
        1. Pericardial aplasia
        2. Ectopia cordis
        3. Sternal aplasia
        4. Anterior diaphragmatic hernia
        5. Abdominal wall defect (omphalocele)

## **Pectus Excavatum** (Fig. 1)

1. The most common chest wall deformity.
2. Approximate incidence is 1 in 400 births although true incidence is unknown.
3. Etiology unknown, more common in Caucasians and over 40% have a family history.
4. May be present at birth or be noted later and may become more severe during the adolescent growth spurt.
5. More frequent in males 4.5:1 vs. females.
6. May be associated with other musculoskeletal abnormalities including scoliosis up to 30% and syndromes including:

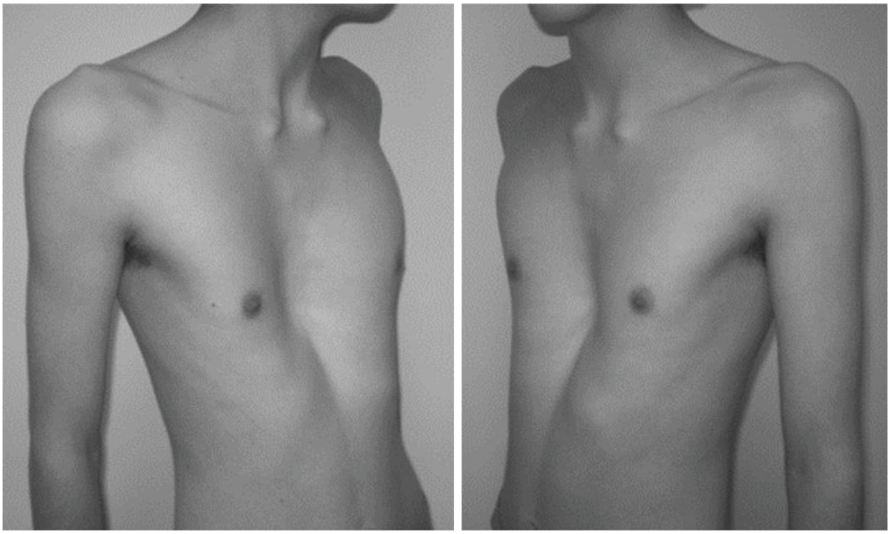

**Fig. 1** Pectus excavatum (Source: DP Croitoru personal files)

   a. Marfan syndrome—earlier presentation and greater severity of defect. Evaluation for aortic root dilation due to a risk of aortic dissection should be done.
   b. Ehlers–Danlos syndrome: collagen defect with hyperextensible joints and loose skin.

7. Pathophysiology: symptoms often do not become manifest until the pubertal growth spurt when the deformity may markedly increase in depth over a short period of time.

   a. Pulmonary—symptoms include shortness of breath and exercise intolerance as well as increased incidence of respiratory infections and asthma. A restrictive defect on pulmonary function testing may be found. Abnormal chest wall motion abnormalities have been noted and are not usually tested. Repair decreases shortness of breath, increases exercise tolerance and oxygen pulse as an estimate of cardiac output.
   b. Cardiac—symptoms include palpitations, occasional syncope and murmurs on examination. Cardiac echocardiogram may demonstrate cardiac compression, mitral valve prolapse, valve insufficiency, and regurgitation as well as possible aortic root dilatation. Repair often relieves mitral valve prolapse and valve insufficiencies and relieves cardiac compression improving cardiac output.
   c. Body image/psychosocial: symptoms include body consciousness when chest is exposed, avoidance of activities exposing chest such as swimming or sports, use of loose clothing to hide defect, teasing in school or sports and behavioral changes including depression, and even suicidal ideation. The

   “cosmetic component” is still often erroneously thought to be the only aspect of the disease ignoring the physiologic aspects of the deformity. When sternal torsion is associated with the deformity over 90% of the time the right side is deeper than the left and in female patients there may be marked pseudo-hypoplasia of the breast making it difficult for clothing in addition to physical symptoms.
   d. Long-term effect—longitudinal studies of unrepaired severe deformities have not been done.

8. Evaluation of the patient with Pectus Excavatum
   a. History including psychosocial symptoms and physical examination—
      i. Chest—including ribs, sternum, clavicles, breath sounds, and heart tones/rhythm, note if the patient has a barrel chest.
      ii. Back—examine for scoliosis and kyphosis as well as shoulders for Sprengel deformity (winged scapula).
      iii. Examine for associated signs of Marfan or Ehlers–Danlos Syndromes: lens abnormality, high arched palate, tall height, long thin fingers, hyper-extensible joints, heart murmur.
   b. Pulmonary Function Testing—evaluate for restrictive defect if severe defects or if contemplating repair; (not required for every patient).
   c. Echocardiography—evaluate for cardiac compression, valve competence, mitral valve prolapse, and aortic root size.
   d. Computed Tomography Chest (non-contrast) or MRI
      i. Calculate Haller Index: At the level of the deepest portion of the deformity measure the internal transverse distance of the thoracic cavity and divide by the antero-posterior distance of the thoracic cavity (from the front of the spine to the back of the sternum or deepest soft tissue impingement) at the same level (Fig. 2). A normal value is 2.5 or less. Haller Index greater than 3.24 is considered a more severe pectus excavatum deformity. The

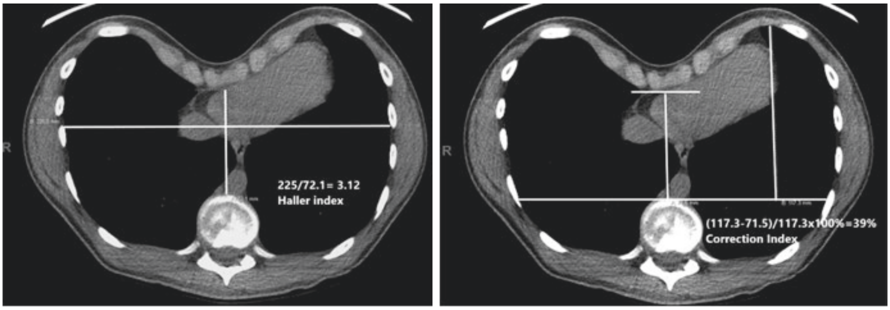

**Fig. 2** Calculation of Haller CT index and CT Correction index. CT scan of patient in Fig. 1 with relatively low Haller Index 3.12 due to barrel chest and high Correction Index 39% (Source: DP Croitoru personal files)

presence of kyphosis on exam producing a barrel chest will alter the calculation of the CT Haller index because the compensatory kyphosis makes the corrected index approach a 1:1 ratio, falsely normalizing the degree of the deformity. This calculation can also be affected by sternal torsion and asymmetry. Unfortunately this value is sometimes required for insurance approval of the repair and may not reach the "required number" due to the presence of a barrel chest.

ii. Calculate correction index: measure the antero-posterior distance from most anterior costal ridge to deepest portion of sternal depression and divide by antero-posterior depth of chest and multiply by 100%. A value of greater than 10% is said to be a significant deformity (Fig. 2). This newer index better discriminates between normal and abnormal pectus excavatum but may also be affected by asymmetry of the chest.

9. Treatment of pectus excavatum

    a. Observation only with no activity restriction is always an option especially for asymptomatic patients since the longitudinal studies on the effect of having a pectus excavatum are lacking. There is no optimal window for repair and operative repairs may be done in adults with caveat that adults have a much longer recovery period and are not as malleable as younger patients.
    b. Non-operative therapy may be attempted with a vacuum bell device (Fig. 3 manufactured by Eckart Klobe (www.tricherbrust.de). This therapy can be successful; however, it requires significant patient compliance and works best for younger malleable patients and therefore the success rate is relatively low.
    c. Operative repair—was often done with open techniques even on very young patients in the past. However, open operation on young patients had a high incidence of recurrence and acquired asphyxiating thoracic dystrophy and is no longer recommended by most surgeons. While there is no upper or lower age limit on performing operative repairs, patients that have entered their

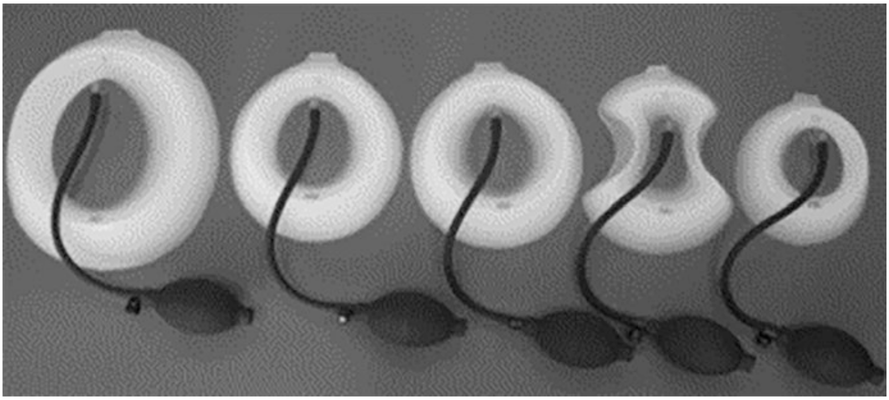

**Fig. 3** Vacuum bell device Eckart Klobe www.tricherbrust.de

pubertal growth spurt and whom are close to achieving skeletal maturity are considered most ideal. Repair of symptomatic pectus excavatum has been shown to reduce shortness of breath, improve exercise tolerance, and reduce compression of the heart.

Operative repair is of two major types.

i. Minimally Invasive Pectus Excavatum Repair (Nuss procedure reported in 1997 after 10 years by Nuss, Kelly, Croitoru et al.) has become the most commonly used method for repair in pediatric and adult patients.

    1. Bilateral lateral thoracic or inframammary incisions are made at the level of the deepest portion of the deformity and depending on the level of the deformity subcutaneous or subpectoralis tunnels are created to the costochondral ridge bilaterally. Utilizing thoracoscopic guidance a dissector is used to create an intrathoracic tunnel from the medial aspect of the costochondral ridge under the sternum and exiting at the medial aspect of the contralateral costochondral ridge to elevate the sternum. A surgical stainless steel bar is curved to the shape of the anticipated corrected thorax and is inserted initially with the curve facing posteriorly through the thoracic tunnel utilizing a chest tube as a sheath to guide to the bar through the tunnel. The bar is flipped 180° to push the sternum into position utilizing the fulcrum of the bilateral costochondral junctions and the underside of the sternum to correct the deformity. In adolescent patients this may require more than 1 bar. A lateral stabilizer is often placed on one side of each bar to help maintain positioning and the bar is fixed to the fascia or around the rib with absorbable sutures. The skin incisions are closed with absorbable sutures. The support bars are removed between 24 and 36 months as an outpatient procedure (Fig. 4).
    2. Perioperative recovery is facilitated by adequate pain management often with a thoracic epidural and patients ambulate no later than the first post-operative day. Recently, cryoablation of the intercostal nerves bilaterally has been shown to decrease length of hospitalization. Hospitalization is from 3 to 5 days with limitations on torsion of the thorax and lying on the side for 7–10 days to prevent possible bar movement and additional pain. A standing posterior-anterior (PA) and lateral chest X-ray for baseline positioning is performed at the time of discharge. Patients are encouraged to walk and perform light activities returning to school after 2 weeks and may return to all activities after 2 months including contact sports.
    3. Complications should be minimal after this operation but have included hemorrhage (<1%), lung and cardiac injury (rare), arrhythmia (rare), pneumothorax (<2%), infection (<1%), effusion, seroma, prolonged or chronic pain, bar dislodgement (<2%), allergy to metal in bar (<1%), and recurrence of the defect (<2%) (this is seen more in patients operated on prior to the adolescent growth spurt).

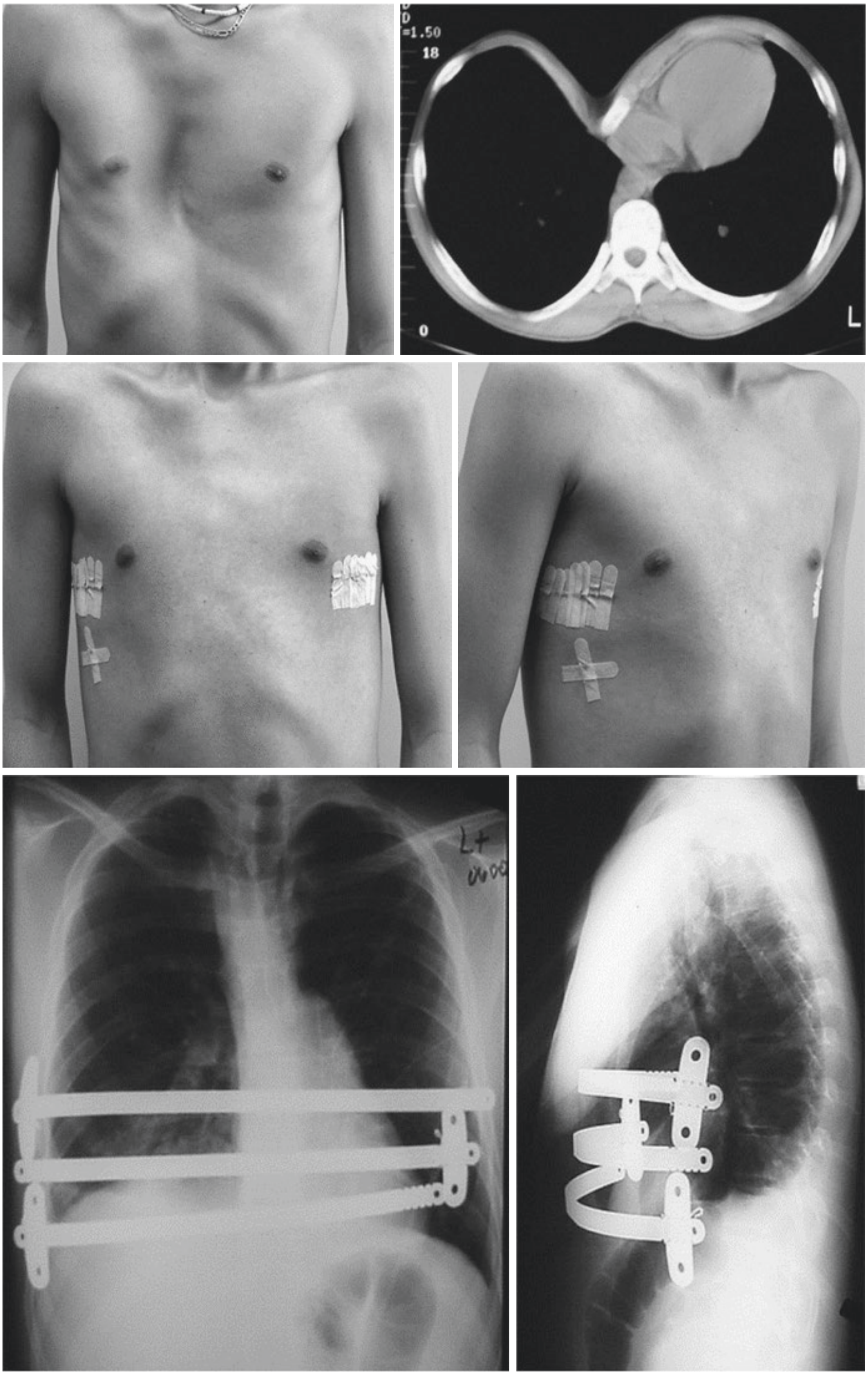

**Fig. 4** A 19-year-old skeletally mature male with severe sternal torsion preop/6 days postop/3 bars (Source: DP Croitoru personal files)

    ii. Open pectus excavatum repair (Ravitch operation): This method has been performed for many years with multiple variations of extent of resection and sub or anterior sternal supports since the initial report in 1949.

        1. A transverse or midline anterior thoracic incision is made and the pectoralis muscles are elevated bilaterally from the sternum and costochondral cartilages. Bilateral subperichondrial resection of the cartilage segments in the area of the deformity are resected (adjusting from ribs 3 to 7 depending on the deformity). A sternal wedge osteotomy is performed and is secured either with sutures or a retrosternal strut secured bilaterally at the 4th rib is placed. The pectoralis flaps are sutured usually with closed suction drains in place and the skin is closed with absorbable sutures.

           If a substernal strut has been placed, it is removed as an outpatient procedure after several months.
        2. Perioperative recovery is facilitated by adequate pain management and patients ambulate no later than the first post-operative day. Hospitalization is from 2 to 5 days with limitations on contact sports for 6 months.
        3. Complications after this operation have included hemorrhage, lung and cardiac injury, arrhythmia, pneumothorax, infection, effusion, seroma, prolonged or chronic pain, recurrence (seen more in patients operated on prior to the adolescent growth spurt), and acquired thoracic dystrophy (seen more in patients operated at very young ages and prior to the adolescent growth spurt).

## Pectus Carinatum (Fig. 5)

1. Second most common chest wall deformity—may be asymmetric.
2. Incidence is much less than pectus excavatum in Caucasians and much more common in patients of Hispanic heritage in South America. True incidence is unknown.
3. Etiology unknown and over 25% have a family history.
4. Usually not present at birth and is noted later and may become more severe during the adolescent growth spurt.
5. More frequent in males 4:1 vs. females.
6. May be associated with other musculoskeletal abnormalities including scoliosis and syndromes including:
    a. Marfan syndrome—earlier presentation and greater severity of defect. Must be evaluated for aortic root dilation due to a risk of aortic dissection.
    b. Ehlers–Danlos syndrome: collagen defect with hyperextensible joints and loose skin.

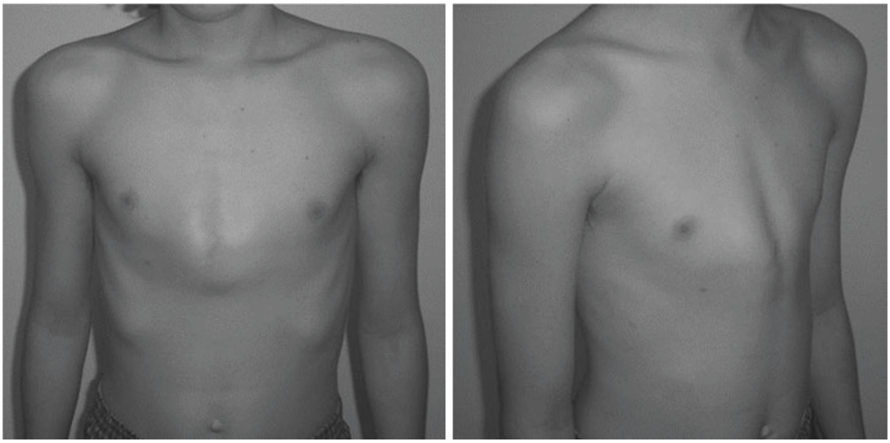

**Fig. 5** Pectus Carinatum (Source: DP Croitoru personal files)

7. Pathophysiology: There are usually no pulmonary or cardiac symptoms with this deformity and musculoskeletal symptoms often do not become manifest until the pubertal growth spurt when the deformity may markedly increase over a short period of time.
    a. Musculoskeletal–-discomfort or pain with lying prone may be associated with this deformity.
    b. Body image/psychosocial: symptoms include body consciousness when chest is exposed, avoidance of activities exposing chest such as swimming or sports, use of loose clothing to hide defect, teasing in school or sports and behavioral changes including depression, and even suicidal ideation. The "cosmetic component" is still often the main component of this deformity.
    c. Long-term effect—longitudinal studies of uncorrected severe deformities have not been done.
8. Evaluation of the patient with Pectus Carinatum
    a. History including psychosocial symptoms and physical examination
        i. Chest—including ribs, sternum, clavicles, breath sounds, and heart tones/rhythm. Note whether the patient has a barrel chest.
        ii. Back—examine for scoliosis and kyphosis as well as shoulders for Sprengel deformity (winged scapula).
        iii. Examine for associated signs of Marfan or Ehler–Danlos Syndromes: lens abnormality, high arched palate, tall height, long thin fingers, hyperextensible joints, heart murmur.
9. Treatment of pectus carinatum
    a. Patients can be treated successfully with non-operative therapy utilizing an orthotic brace which places pressure on the protrusion of the sternum or verti-

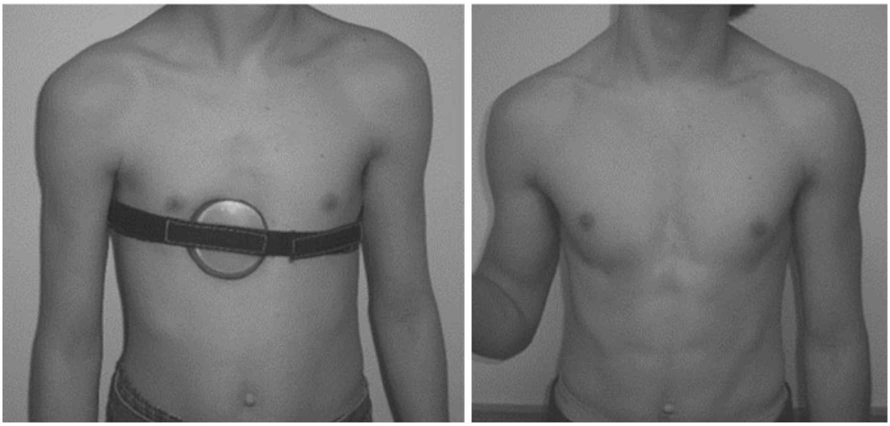

**Fig. 6** Orthotic treatment of patient seen in Fig. 5 and final result (Source: DP Croitoru personal files)

cal ridge. Success is very dependent on patient compliance and flexibility of the chest wall and therefore is more successful during the adolescent growth spurt (Fig. 6).

b. Operative repair may be done by open (Ravitch technique) and has also been done utilizing a modification of the minimally invasive pectus excavatum repair (Nuss) technique placing the bar anterior to the sternum with fixation of stabilizers to the ribs bilaterally.

## Additional Notes

Chest wall deformities have been of interest to pediatric surgeons since many of these deformities are noted at or soon after birth. The most common of these deformities pectus excavatum and pectus carinatum may progress and increase in severity during accelerated growth and adolescence. Dr. Mark Ravitch who had an interest in chest wall deformities modified the technique of Dr. Ernst Ferdinand Sauerbruch from Germany who performed bilateral cartilage resections and sternal osteotomy and in his paper from 1949 recommended not performing the external traction that Sauerbruch and Ravitch himself had initially supported. Ravitch also had the belief that short diaphragmatic ligaments helped to produce the deformity and advocated radical dissection of the diaphragmatic and rectus muscle attachments to the sternum in addition to the bilateral cartilage resections and sternal osteotomy. Dr. Kenneth Welch in Boston did not perform quite so radical a release of the sternum, however, without the use of external traction recurrence was frequent until the addition of internal fixation which was introduced in 1961 by Adkins. This open technique was the main procedure for repair of pectus excavatum until

Dr. Donald Nuss at the American Pediatric Surgical Association APSA meeting in 1997 reported 10 years of experience with a minimally invasive pectus excavatum repair utilizing internal bracing to remodel the chest wall into a normal configuration. This technique has become the primary technique for repair of pectus excavatum in both children and adults. The open (Ravitch) repair is still utilized and is no longer utilized in very young children as it was recommended in the past and can also achieve very good results.

In general, unless patients are having very severe symptoms, operation should be delayed until the patient is entering into their accelerated adolescent growth spurt confirming that the deformity is in fact increasing and also at a time when the chest wall is still malleable and more amenable to repair. Repair of this deformity is an elective operation and because the longitudinal history of uncorrected pectus excavatum is not known, there are no absolute indications that repair is necessary and it may be delayed and performed in adults. The caveat to performing these corrective operations on older patients is that their recovery as would be expected is more prolonged.

## Study Questions

1) Pectus excavatum repair should be recommended:

   A. As soon as it is diagnosed and before age 5 to prevent it from progressing
   B. When the patient has reached skeletal maturity
   C. When the Haller index is 3.2 or correction index is >10%
   D. Electively if the patient is symptomatic and is interested in having repair
   E. C and D.

Answer: Answer: (E.) The repair of pectus excavatum is almost always a totally elective procedure and history has shown that performing the procedure on younger patients while reportedly easier due to the fact that younger children are more malleable early repair does not prevent the deformity from progressing and may in fact lead to the development of acquired thoracic dystrophy which can be a devastating sequela to operation at an early age. Repairs of the deformity may be done after reaching skeletal maturity and can be performed in adults with good success, however patients that have reached skeletal maturity are not as malleable and the recovery period is therefore more prolonged. Consideration to undergo repair should include findings that the patient is symptomatic and in cases where they have a Haller index greater than 3.2 or a correction index greater than 10%. There are patients that have an index greater than 3.2 and have minimal symptoms due to adaptation from having the deformity since a young age. There are no absolute indications for urgent surgical repair and because the longitudinal history is unknown patients may live with this deformity.

2) Pectus carinatum is:

   A. More commonly manifest in South America and in patients of Hispanic descent
   B. A surgical emergency requiring repair as soon as possible
   C. Can often be corrected with orthotic bracing
   D. Is always associated with Marfan syndrome
   E. A and C

Answer: (E.) Pectus carinatum may be associated with Marfan syndrome in a small percentage of cases as can the deformity of pectus excavatum. Both of the two most common chest wall deformities can be associated with Ehlers–Danlos syndrome as well. Surgical repair of pectus carinatum is never absolutely required and this deformity may be corrected with orthotic bracing if patients are compliant with treatment. It is more commonly seen in patients of Hispanic descent and is therefore more common in South America.

3) The most common symptoms and findings associated with pectus excavatum are:

   A. Shortness of breath, exercise intolerance, and scoliosis
   B. Palpitations, asthma, Raynaud's syndrome
   C. Chest wall motion abnormalities, hyperflexibility, and mitral valve prolapse
   D. Marfan syndrome, Ehlers–Danlos syndrome, and aortic root dilatation

Answer: (A.) While all of the symptoms and findings in the answers above have been noted in association with pectus excavatum, the most common presenting symptoms and findings include shortness of breath, exercise intolerance, and the associated finding of scoliosis.

## Further Reading

Abramson H, D'Agostino J, Wuscovi S. A 5-year experience with a minimally invasive technique for pectus carinatum repair. J Pediatr Surg. 2009;44:118–24.

Croitoru DP, Kelly RE, Goretsky MJ, et al. Experience and modification update for the minimally invasive Nuss technique for pectus excavatum repair in 303 patients. J Pediatr Surg. 2002;37(3):437–45.

Daeman JH, de Loos ER, Vissers YLJ, et al. Intercostal nerve cryoablation versus thoracic epidural for postoperative analgesia following pectus excavatum repair: a systematic review and meta-analysis. Interact Cardiovasc Thorac Surg. 2020;31(4):486–98.

Graves C, Moyer J, Zobel M, et al. Intraoperative intercostal nerve ablation during the Nuss procedure reduces length of stay and opioid requirement: a randomized clinical trial. J Pediatr Surg. 2019;54(11):2250–6.

Haje SA, Bowen JR. Preliminary results of orthotic treatment of pectus deformities in children and adolescents. J Pediatr Orthop. 1992;12:795–800.

Haller JA Jr, Loughlin GM. Cardiorespiratory function is significantly improved following corrective surgery for severe pectus excavatum. Proposed treatment guidelines. J Cardiovasc Surg (Torino). 2000;41(1):125–30.

Haller JA Jr, Kramer SS, Lietman SA. Use of CT scans in selection of patients for pectus excavatum surgery: a preliminary report. J Pediatr Surg. 1987;22:904–8.

Kaguraoka H, Ohnuki T, Itaoka T, et al. Degree of severity of pectus excavatum and pulmonary function in preoperative and postoperative periods. J Thorac Cardiovasc Surg. 1992;104(5):1483–8.

Kelly RE, Goretsky MJ, Obermeyer R, et al. Twenty-one years of experience with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. Ann Surg. 2010;252(6):1072–81.

Kowalewski J, Brocki M, Dryjanski T, et al. Pectus excavatum: increase of right ventricular systolic, diastolic, and stroke volumes after surgical repair. J Thorac Cardiovasc Surg. 1999;118(1):87–92.

Lawson ML, Mellins R, Tabangin M, et al. Impact of pectus excavatum on pulmonary function before and after repair with the Nuss procedure. J Pediatr Surg. 2005;40(1):174–80.

Lawson ML, Mellins RB, Paulson JF, et al. Increasing severity of pectus excavatum is associated with reduced pulmonary function. J Pediatr. 2011;159(2):256–61.

Mielke CH, Winter RB. Pectus carinatum successfully treated with bracing: a case report. Int Orthop. 1993;17:350–2.

Nuss D, Kelly RE, Croitoru DP, et al. A 10-year review of a minimally invasive technique for the correction of pectus excavatum. J Pediatr Surg. 1998;33:545–52.

Ravitch MM. The operative treatment of pectus excavatum. Ann Surg. 1949;129:429–44.

Schier F, Bahr M, Klobe E. The vacuum chest wall lifter: an innovative, nonsurgical addition to the management of pectus excavatum. J Pediatr Surg. 2005;40:496–500.

Shamberger RC, Welch KJ. Surgical correction of pectus carinatum. J Pediatr Surg. 1987;22:48–53.

St. Peter SA, Juang D, Garey CL, et al. A nover measure for pectus excavatum: the correction index. J Pediatr Surg. 2011;46(12):2270–3.

# Inflammatory Bowel Disease

**Sasha Slipak and Christopher P. Coppola**

**Key Points**

1) Inflammatory bowel disease is caused by an abnormal immune response to the intestinal mucosa resulting in inflammation, ulcers, gastrointestinal symptoms, and in severe cases, obstruction and perforation.
2) Crohn's disease cannot be definitively cured; the goal of therapy is to keep the disease in remission through pharmaceutical treatment and avoid loss of length of intestine through operative therapy.
3) In ulcerative colitis, there is a cumulative risk of colon cancer developing after adolescence so total colectomy with J-pouch reconstruction is recommended in all patients.

## Introduction

Inflammatory bowel disease (IBD) encompasses two similar but distinct disease processes that cause idiopathic inflammation of the gastrointestinal tract [1]. IBD disease process may involve an altered immune system, genetic predisposition, and environmental factors to varying degrees. Crohn's disease (CD) has no known cure but does have complicated treatment options which span medical and surgical

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expertise. Total colectomy is the only option for cure in ulcerative colitis (UC). Treatment is best offered in a multidisciplinary fashion with the goal of therapy to control disease and maintain patients in remission.

## Crohn's Disease

1. Pathophysiology
    a. An abnormal inflammatory response to the intestinal mucosa leads to ulceration (aphthous ulcers) that form in linear fashion. The surrounding mucosa becomes edematous and forms a distinctive appearance of "cobblestone" [2].
        i. Histologically the bowel wall reveals neutrophilic as well as eosinophilic inflammatory cells.
        ii. In nearly half the cases, a non-caseating granuloma will be seen and is pathognomonic to the CD.
    b. As the inflammation progress it affects all the layers of the bowel and will lead to the hallmark "creeping fat" as the mesentery surrounding the affected bowel thickens and proliferates.
        i. Submucosa is edematous and fibrotic.
    c. If the inflammation progresses, it may lead transmural edema with sinus creation and fistula formation. Chronic inflammation may form strictures and rarely perforation.
    d. Crohn's disease may involve the entire gastrointestinal tract and may be difficult to distinguish from UC. Most commonly the ileum and cecum are affected together. The disease can be isolated just to the small bowel, most likely the ileum, or more rarely isolated to the colon.
2. Clinical features
    a. Most common presenting signs and symptoms:
        i. Crampy abdominal pain
            1. Local peritonitis, mass in RLQ, and fever may indicate perforation.
        ii. Weight loss/failure to thrive; it is common to see a child "fall off" the growth chart
            1. Decreased appetite
            2. Growth and sexual maturation delay.
        iii. Diarrhea and/or multiple bowel movements
            1. Occult blood present in stool. Frank blood may not be present.

b. Other signs and symptoms:
    i. Perianal disease
        1. Anal fissures
        2. Perianal fistulas
        3. Perianal abscess
    ii. Erythema nodosum
    iii. Arthritis and/or arthralgia
    iv. Episcleritis and uveitis
    v. Nephrolithiasis
    vi. Hepatobiliary symptoms
        1. Primary sclerosing cholangitis
        2. Cholelithiasis
        3. Hepatitis
        4. Steatosis.

3. Diagnosis
    a. History and physical: Crohn's disease affects multiple body systems so a thorough head to toe physical examination is very important.
        i. Trend child's weight and recent complaints/hospitalizations.
        ii. Family history.
        iii. Special attention to the abdominal exam which may reveal a subtle mass in the right lower quadrant.
    b. Lab work
        i. Complete blood count (CBC): looking for anemia.
        ii. Basic metabolic panel (BMP): electrolyte disturbance.
        iii. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP): inflammatory markers may be elevated in acute flare-ups.
        iv. Nutritional studies and hepatic function: may be abnormal.
        v. IgA and IgG anti-saccharomyces cerevisiae antibodies are highly specific for CD in children.
        vi. Send stool to be checked for blood and infectious agents.
    c. Rectal exam
        i. In a younger child or a very anxious patient examination under anesthesia may be advantageous.
            1. Anorectal endoscopic ultrasound which may help in detecting an abscess.
    d. Imaging
        i. Upper gastrointestinal (UGI) contrast study with small bowel follow-through (SBFT) may show strictures.

    ii. Computed tomography (CT) imaging may show mucosal nodularity, area of inflammation, abscess, creeping fat.
    iii. Magnetic resonance imaging (MRI) with gadolinium enhancement is sensitive to identifying proximal small bowel disease.

  e. Endoscopy

    i. Ileocolonoscopy: be cautious if there is concern for perforation.

      1. Direct observation

        a. Skip lesions: cobblestone appearance
        b. Aphthous ulcers
        c. Inflamed terminal ileum

      2. Terminal ileum biopsy.

    ii. Upper endoscopy: may have lesions in the upper gastrointestinal tract as well.
    iii. Capsule endoscopy may have a role in certain patients.

4. Initial management

  a. If no sign of perforation or obstruction, then medical treatment is first line

    i. Intravenous fluids and correction of electrolyte abnormalities.
    ii. Mesalamine (an aminosalicylate) is the first-line therapy for mild to moderate active CD.
    iii. Immunosuppressant medication

      1. Steroids
      2. Azathioprine
      3. 6-mercaptopurine.

  b. Concern for perforation requires exploratory laparotomy and resection

    i. May need a temporary ostomy while the inflammation subsides.
    ii. Crohn's disease with perforation may mimic other cause of acute abdomen such as perforated appendicitis.

  c. Obstruction requires operative exploration and as minimal resection as possible.
  d. Intraabdominal abscesses are best treated with image guided drainage and antibiotics.
  e. Fistulae between bladder, uterus, and other segment of intestine are treated with limited resection.
  f. Perianal abscess and complex fistulas are treated surgically with the least disruption to the sphincter as possible.

    i. Complex fistulas may require a diverting colostomy.
    ii. Use a noncutting seton: avoid reactive sutures/seton as this may lead to further inflammatory response.
    iii. Fibrin glue may also be attempted.

5. Treatment
    a. Multidisciplinary team approach
        i. Gastroenterologist
        ii. Pediatrician
        iii. Pediatric surgeon
        iv. Nutritionist
        v. Social worker
    b. Maintaining patient in remission is goal of treatment [3].
        i. Infliximab
            1. Test for latent tuberculosis before infliximab therapy is started.
        ii. Metronidazole and ciprofloxacin in mild to moderate disease
            1. May help delay recurrence at surgical anastomosis
            2. Aid in healing of perianal fistula.
        iii. Dietary change/enteral therapy
            1. Total liquid diet
            2. Exclusive enteral nutrition
                a. Enteral formulas with transforming growth factor-β (TGF-β) and omega-3 fatty acids.
        iv. Surgical treatment
            1. If medical therapy fails to control the flare-up, then resection of the affected area is indicated.
            2. Ileocecectomy is the treatment of choice for patients with isolated ileocecal disease.
            3. Stricturoplasty for obstruction or near obstructions
                a. Heineke–Mikulicz for 1–10 cm strictures
                b. Finney stricturoplasty for longer strictures
                c. Michelassi technique—for longer strictures (side-to-side isoperistaltic stricturoplasty).
6. Outcome
    a. No cure for Crohn’s disease is available.
    b. Remission is goal.
    c. Minimize bowel resection if surgical intervention is required.
    d. Colorectal and small bowel cancer is increased in CD
        i. Screening for colorectal cancer should start 10 years after start of symptoms.

## Ulcerative Colitis

1. Pathophysiology
    a. Chronic inflammatory response confined to mucosa and does not extend into the muscularis propria. The inflammation usually involves the rectum and extends proximally in a contiguous fashion without skip lesions.
        i. Microscopically: crypt abscesses and mucosal ulceration.
        ii. Undermining of mucosa by ulcerations leads to mucosal bridging and characteristic pseudopolyp formation in circumferential pattern.
    b. The acute phase of the disease significantly affects the colon.
        i. Distention, decreased peristalsis, muscularis propria thins and bleeds.
        ii. If the acute phase is not treated, it may progress to toxic megacolon: severe colonic dilatation, peritonitis, and even perforation.
    c. In chronic disease a characteristic lead pipe appearance is observed on imaging
        i. The muscularis propria thickens and haustral folds are flattened with decreased peristalsis.
        ii. Mucosal surface becomes atrophic and may develop dysplasia.
        iii. Mesentery may shorten and serosal surface increase in vascularity as well as adipose tissue.
2. Clinical features: presentation can range from mild to severe
    a. Most common presenting signs and symptoms:
        i. Multiple daily loose bowel movement
            1. The diarrhea may have blood, mucus, and pus.
            2. Significant disease may have frank blood per rectum necessitating blood transfusions.
        ii. Abdominal pain: intermittent and crampy
            1. Decreased appetite or anorexia
            2. Rapid weight loss in severe disease.
        iii. Tenesmus.
        iv. In severe cases patient may present in sepsis: toxic megacolon.
        v. Aphthous stomatitis often present during active disease.
    b. Other signs and symptoms
        i. Anemia
            1. Secondary to bloody diarrhea
            2. Anemia of chronic disease
        ii. Fevers
        iii. Hemorrhoids

        iv. Erythema nodosum
        v. Arthralgia
        vi. Pyoderma gangrenosum
            1. Most common in the lower extremities and occasionally the trunk
        vii. Uveitis
        viii. Nephrolithiasis
        ix. Delayed sexual maturation
        x. Primary Sclerosing Cholangitis.

3. Diagnosis
    a. History and physical: due to the multiple body systems involved a thorough exam is very important.
        i. Signs of anemia
        ii. Signs of dehydration
        iii. Signs of sepsis and/or shock in severe cases.
    b. Lab work
        i. CBC: looking for anemia.
        ii. BMP: electrolyte disturbance with persistent diarrhea.
        iii. ESR and CRP: inflammatory markers may be elevated in acute phase.
        iv. Nutritional studies
            1. Hypoalbuminemia secondary to protein loss from ulcerated colonic mucosa.
        v. Send stool to be checked for leukocytosis, blood, and for infectious etiology.
            1. Clostridium difficile infection is possible and can significantly complicate the diagnosis.
    c. Rectal exam
        i. External hemorrhoids are common.
        ii. Absence of fistulas, fissures, and abscess. If seen may suggest CD.
    d. Imaging
        i. UGI contrast study with SBFT helps to identify if there is small bowel disease which would suggest CD rather than UC.
        ii. Abdominal plain film X-ray
            1. If toxic megacolon suspected—get plain film first.
                a. Quickly identify if free air is present.
        iii. CT imaging may show mucosal thickening of the large bowel with sparing of the small bowel.
        iv. MRI with gadolinium enhancement is sensitive to identifying proximal small bowel disease and less sensitive for large bowel: may be a better

choice due to lack of ionizing radiation and the multiple imaging the child will most likely receive in their lifetime.

v. Barium enema (BE): Not commonly used due to concern of barium stimulating acute manifestations of the disease.
    1. Classic lead pipe appearance due to loss of the haustral folds.
    2. Pseudopolyps may be visible.

e. Endoscopy
    i. Ileocolonoscopy may be very helpful in establishing diagnosis between CD and UC.
    ii. Flexible sigmoidoscopy with biopsy
        1. Direct observation
            a. friable and edematous colonic mucosa with a thin, purulent exudate.
    iii. Upper endoscopy: Use if unsure between UC and CD.

4. Initial management
    a. Toxic megacolon presentation especially with perforation will require immediate surgical intervention
        i. Total colectomy with end ileostomy.
    b. Severe rectal bleeding and severe symptoms refractory to medical treatment also require urgent surgical intervention.
    c. If no sign of perforation or shock:
        i. 5-aminosalicylic acid, 5-ASA is first line of therapy in mild to moderate disease.
        ii. IV fluids and correction of electrolyte abnormalities.
        iii. In severe disease IV steroids.
        iv. If disease refractory to other treatment options, Cyclosporine may be given.

5. Treatment
    a. Multidisciplinary team approach
        i. Gastroenterologist
        ii. Pediatrician
        iii. Pediatric surgeon
        iv. Nutritionist
        v. Social worker.
    b. Maintaining patient in remission or potential cure is goal of care
        i. Long-term steroids and other immunosuppressants lead to higher complication rates after surgery

        1. The sooner the colectomy is done the less likely complications to follow.
        2. Due to the potential cure of disease elective colectomy should be considered.

    ii. Total colectomy with ileoanal pouch procedure is now the preferred surgical option for elective colectomy secondary to UC

        1. Distal rectal mucosectomy (lower 4 cm) performed in order to spare the pelvic nerves and sphincter muscles.
        2. J-shaped pouch is most common. S-shaped pouch is also commonly constructed. Key is to keep pouch length approximately 8 cm in length.
        3. A temporary loop ileostomy is created to protect the anastomosis. This is associated with decreased risk of failure [4].
        4. A water-soluble contrast enema is performed within 2 months to ensure that the ileal reservoir has no leaks or sinus tracts at which time the loop ileostomy is usually reversed.
        5. Significantly increased stooling frequency is common which typically gets better in several months.

            a. Multiple stools each day is common lifelong.

                i. Bulking agents and anti-diarrhea medication may help to decrease frequency.

            b. Pouchitis is most frequent complication.

                i. Crampy lower abdominal pain and increased frequency of stooling, often with watery diarrhea.
                ii. Initiate metronidazole or ciprofloxacin. Barium enema to confirm pouch is open. Consider daily pouch washout with tap water. If needed diverting ileostomy.
                iii. Probiotics may decrease the recurrence of pouchitis.

            c. In cases of pull-through failures a permanent ileostomy is required.

6. Outcome

    a. Medical management of UC with goal of remission.
    b. Surgical treatment with goal of cure

        i. Patient and family need to be well informed of what to expect after surgery and the possible complications.

    c. Colorectal cancer is increased in UC

        i. Three percent risk of colorectal cancer in first decade with 10–15% increased risk each subsequent decade after the diagnosis.
        ii. Evidence of dysplasia on biopsy are at increased risk of developing a carcinoma.
        iii. Screening should start 10 years after diagnosis.

            1. May consider sooner if dysplasia initially seen on biopsy.

## Indeterminate Colitis

1. Pathophysiology: Patients with overlapping symptoms of Crohn's disease and ulcerative colitis based on biopsy. May represent more severe cases which makes diagnosis difficult.
   i. May represent up to 15% of cases.
2. Treatment: Disease confined to the colon should be treated with total colectomy with ileal reservoir.
   i. The fact that some may eventually be diagnosed with Crohn's disease should not be a contraindication to surgical treatment.

## Additional Notes

Inflammatory bowel disease (IBD) encompasses a spectrum of disorders which can be divided into two broad subcategories: Ulcerative colitis (UC) and Crohn's disease (CD). These are relapsing, inflammatory conditions of the gastrointestinal tract with distinct yet overlapping clinical and pathological features. While the etiology of these two inflammatory bowel diseases is unknown, current data suggests that an environmental trigger causes disease in a host genetically predisposed due to intrinsically impaired immunity. Some combination of host factors (family history, ethnicity, genetic predisposition, immunity), gut factors (microbiome, epithelial barrier, pathogen recognition, autophagy, T cell mediated response), and environmental factors (geographic location, smoking, NSAIDs, bacterial/viral pathogens) lead to the development of mucosal inflammation and result in the development of IBD.

The medical and surgical management of Crohn's and UC is in many aspects similar between children and adults. In children, IBD may manifest at any age but occurs most commonly between late adolescence and early adulthood. There also appears to be a slight female predominance. Unlike adults, children with IBD tend to present differently. There is often a longer delay to establishing a definitive diagnosis in children as they present with systemic symptoms including fever of unknown origin, arthritis, chronic microcytic anemia, or even delayed growth or development. This may precede any overt gastrointestinal manifestations.

In children presenting with Crohn's disease, 33% have disease limited to the small intestine, 45% have disease of the small bowel and colon, 20% have disease of the colon alone, and roughly 33% have perianal disease. This, however, is rarely isolated. In addition, most of these patients will also have some oral ulcers which are rarely severe. As in adults, this disease has no known cure, but does have an array of treatment options including both medical and surgical. A special consideration in children is the long-term effect of medical management on growth, bone health, and social development. The risk of years of toxic therapy must be balanced with the dangers of uncontrolled inflammation and its long-term sequelae (fibrosis, fistulas, cancer).

Ulcerative colitis in children tends to present with extra-intestinal manifestations, most commonly polyarthritis involving large joints. This rarely results in joint destruction. Unlike Crohn's disease, in UC there is a parallel in the severity of inflammation and intestinal involvement. This gives the option of cure for not only the colitis, but the extra-intestinal disease processes as well. This may be achieved through total proctocolectomy which is often followed by a restorative ileal pouch procedure. Of note, as in adults, any bile duct pathology does not seem to resolve following curative surgery. Regardless of the disease process, treatment is best offered in a multidisciplinary fashion with the goal of therapy to control disease and maintain patients in remission.

## Study Questions

1) Question 1: Which statement is true about Crohn's disease?
   a. There is a cumulative risk of colon cancer so all patients should undergo colectomy.
   b. When patients are admitted for gastrointestinal symptoms, resection of affected intestine is the treatment of choice.
   c. The inflammation in Crohn's disease is characterized by aphthous ulcers that form in a linear fashion.
   d. Crohn's disease does not lead to perforation.

Answer to Question 1: (c.) The inflammation in Crohn's disease is characterized by aphthous ulcers that form in a linear fashion. Eventually increases in inflammatory changes will lead to a cobblestone appearance of the mucosa. The inflammation affects all layers of the bowel wall and results in creeping fat appearance of the external surface of the bowel. This transmural inflammation can lead to obstruction and perforation. It is ulcerative colitis that has a cumulative risk of colon cancer, so colectomy is recommended in ulcerative colitis.

2) Question 2: Which of the following is an extracolonic sign of ulcerative colitis?
   a. Erythema nodosum
   b. Pyoderma gangrenosum
   c. Uveitis
   d. All of the above

Answer to Question 2: (d.) All of the above. Ulcerative Colitis is an immune mediated inflammatory condition of the intestine limited to the mucosa. The autoimmune process causes malabsorption and it can be associated with many other symptoms, such as erythema nodosum, arthralgia, pyoderma gangrenosum, uveitis, nephrolithiasis, delayed sexual maturation, primary sclerosing cholangitis, anemia, fevers, and hemorrhoids.

3) Question 3: Which is the preferred operation to prevent colon cancer in individuals with ulcerative colitis?

   a. Left hemicolectomy.
   b. Total colectomy and proctectomy with ileal J-pouch reconstruction
   c. Limited resection of symptomatic regions
   d. Ileostomy and rectal Hartmann's pouch

Answer to Question 3: (b.) Total colectomy and proctectomy with ileal J-pouch reconstruction. All patients with ulcerative colitis have a cumulative risk of developing colon cancer. For this reason, total colectomy and proctectomy is recommended in all patients once they reach adolescence. The preferred reconstruction is ileal J-pouch with ileal–anal anastomosis. Partial colonic resections will leave tissue at risk for dysplasia and cancer. Although some patients will require permanent ileostomy because they cannot achieve a functioning J-pouch, the majority will have a good quality of life with a functional J-pouch after a period of recovery, healing, and adaptation (Fig. 1).

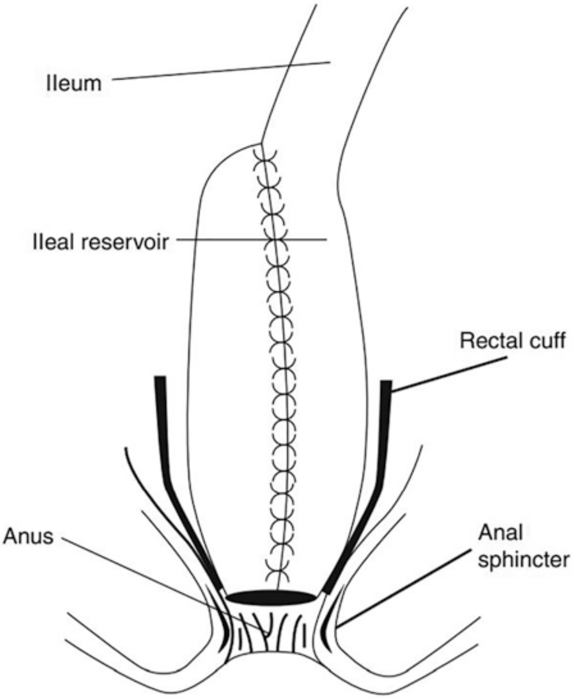

**Fig. 1** J-pouch with ileoanal anastomosis. After proctocolectomy for ulcerative colitis, a J-pouch is created, and an anastomosis made to the rectal cuff creating an ileoanal reservoir for stool. Usually, a protective diverting loop ileostomy is created proximal to the J-pouch to allow it to heal (Adapted from: National Digestive Diseases Clearinghouse, National Institutes of Health, NIH, May 2012. http://digestive.niddk.nih.gov/ddiseases/pubs/dictionary/e-k.aspx. Downloaded on 6 Jan 2014, Used with permission, Image in public domain)

## References

1. National Institute of Diabetes and Digestive and Kidney Diseases. Crohn's disease. Washington, DC: National Institutes of Health, US Department of Health and Human Services. 2021. https://www.niddk.nih.gov/health-information/digestive-diseases/crohns-disease. Accessed 26 Nov 2021.
2. North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition. Differentiating ulcerative colitis from Crohn disease in children and young adults: report of a Working Group of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition and the Crohn's and Colitis Foundation of America. J Pediatr Gastroenterol Nutr. 2007;44(5):653–74.
3. Smith RK, Holubar SD. Medical therapy for Crohn's disease. In: Steele SR, Hull TL, Hyman N, Maykel JA, Read TE, Whitlow CB, editors. The ASCRS textbook of colon and rectal surgery. Cham: Springer; 2022.
4. Westberg K, Olen O, Soderling J, Bengtsson J, Ludvigsson JF, Everhov AH, Myrelid P, Nordenvall C. Primary versus staged reconstruction and risk of surgical failure in patients with ulcerative colitis: a nation-wide cohort study. Inflamm Bowel Dis. 2021; https://doi.org/10.1093/ibd/izab263.

# Multiple Endocrine Neoplasia Syndromes

**Rebekah Campbell and Melissa Mao**

**Key Points**

1) There are three types of Multiple Endocrine Neoplasia (MEN): Type 1 is due to a Menin mutation, and Type 2A and Type 2B are due to Ret mutations.
2) MEN Type 1 has a high penetrance for parathyroid hyperplasia.
3) One-third of patients with gastrinomas have MEN Type 1.
4) MEN Type 2A and 2B are characterized by medullary thyroid carcinoma and pheochromocytomas.

## Introduction

Multiple Endocrine Neoplasia (MEN) Type 1 was discovered in 1954 when a previously reported syndrome of pituitary, parathyroid, and pancreatic tumors was discovered to be inherited by a dominant trait. The earliest report was in 1903 of a patient with a pituitary adenoma causing acromegaly and three enlarged parathyroid glands. In the 1960s, MEN Type 2A and Type 2B were described, and the phrase “multiple endocrine neoplasia” was formed.

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## Outline

I. Multiple Endocrine Neoplasia (MEN) Type 1

  A. Epidemiology:

    1. Autosomal dominant mutation in the gene, Menin.
    2. Menin acts as a tumor suppressor.
    3. Affects 1 in 30,000 people in the USA.
    4. Defined by possessing two of three major syndromes listed below.
    5. One-third of MEN related deaths are caused by MEN Type 1-associated malignancies.

  B. Syndrome

    1. Pituitary Tumors

      a. 10–60% of patients with MEN type 1
      b. Most commonly a prolactinoma (20%)
      c. Can also be Adrenocorticotropic Hormone (ACTH)-secreting or Growth Hormone secreting
      d. Diagnose with brain Magnetic Resonance Imaging (MRI) and serum prolactin levels
      e. Treat with dopamine agonists
      f. Monitor with prolactin levels yearly and MRI brain every 3–5 years

    2. Primary Hyperparathyroidism

      a. Leads to multigland parathyroid hyperplasia. Compare this to the general population, where 80% of primary hyperparathyroidism is due to adenomas
      b. High penetrance rate, and therefore 90% of MEN Type 1 patients get parathyroid hyperplasia by age 40, and nearly 100% by age 50
      c. Diagnosis: elevated serum calcium and parathyroid hormone levels
      d. Surgical Treatment:

        i. Subtotal parathyroidectomy (typically 3 1/2 gland excision) with cervical thymectomy
        ii. Total parathyroidectomy with autotransplantation of 1/2 gland with cervical thymectomy (most commonly used site is the sternocleidomastoid muscle)

      e. Monitor with calcium and parathyroid hormone levels
      f. Recurrence is common and may require further resections

    3. Pancreatic Neuroendocrine Tumors (PNET)

      a. Most common overall: non-functioning PNET
      b. Most common functioning PNET: Gastrinoma (~40%)

        i. If found to have gastrinoma, evaluate for MEN Type 1. The Menin mutation is present in approximately 1/3 of patients with gastrinoma
        ii. Symptoms
            a.) Intractable peptic ulcers due to increased gastrin secretion (Zollinger–Ellison Syndrome)
            b.) Reflux esophagitis
            c.) Chronic diarrhea
        iii. Diagnosis:
            a.) Upper endoscopy: confirm peptic ulcer disease
            b.) Fasting serum gastrin > 10× upper limit of normal AND gastric pH < 2
            c.) If fasting serum gastrin < 10× upper limit of normal, need confirmatory tests with secretin stimulation and/or basal acid output
            d.) Imaging:
                i.) Dotatate positron emission tomography (PET) or somatostatin scintigraphy
                ii.) Look for multiple small tumors in the gastrinoma triangle: Junction of cystic duct and common bile duct, junction of head and neck of pancreas, junction of second and third parts of duodenum
        iv. Treatment
            a.) Parathyroidectomy—correction of hypercalcemia can improve symptoms
            b.) If symptoms are well controlled—medical management with proton pump inhibitors
            c.) Surgical Treatment:
                i.) May include mucosal resection of duodenum along with distal pancreatectomy.
                ii.) Incredibly difficult to remove all tumors surgically, and this should be used primarily as a debulking procedure.
        v. Monitor with gastrin levels and dotatate-PET or somatostatin scintigraphy
    c. Other functioning PNETs: insulinoma (~10%), glucagonoma, vasoactive intestinal peptide tumor (also known as VIPoma)

4. Additional Manifestations
    a. Lipomas
    b. Non-functioning adrenal cortical tumors

        c. Facial angiofibromas
        d. Rare manifestations: pheochromocytoma, carcinoid tumors

    C. Diagnosis

        1. Most common de novo presentation is recurrent hyperparathyroidism in middle age

            a. Patient has history of removal of one parathyroid adenoma in their 20s.
            b. In reality, the patient has parathyroid hyperplasia, not parathyroid adenoma.

        2. The second most common de novo presentation (25%) is anterior pituitary adenoma.
        3. Screening recommendations for patients who have family history of MEN Type 1:

            a. Genetic testing for Menin mutation.
            b. At age 5: fasting glucose, insulin, prolactin, Insulin-like growth factor (IGLF)-I, brain MRI.
            c. At age 8: calcium and PTH levels.
            d. At age 20: gastrin, secretin-stimulated gastrin, chromogranin-A, glucagon, proinsulin, and computed tomography (CT), dotatate-PET, or somatostatin scintigraphy.

II. MEN Type 2A and 2B

    A. Epidemiology:

        1. Autosomal dominant mutation in the gene, Ret
        2. Ret is a proto-oncogene
        3. Affects 1 in 35,000 people in the USA

    B. Syndrome

        1. Medullary Thyroid Carcinoma

            a. Most common cause of death in MEN 2A
            b. Manifests in 90% of patients
            c. Surgical Treatment: prophylactic total thyroidectomy and central neck dissection (all lymphatic tissue between sternal notch and hyoid bone, and between the jugular veins)

                i. Known MEN 2A patients—perform by age 1–5 years old.
                ii. If discovered de novo, after age 10–15, include ipsilateral modified radical neck dissection on affected side due to high incidence of lymph node metastasis.
                iii. If discovered de novo, check calcitonin level, and look for distant metastasis. If distant metastases are present, then do not perform thyroidectomy or lymph node dissections.

            d. Monitor with calcitonin and carcinoembryonic antigen (CEA) levels, and with neck ultrasounds yearly

2. Pheochromocytoma
    a. This must be treated first before any neck surgery for medullary thyroid cancer
    b. Manifests in 50% of these patients
    c. Diagnosis:
        i. Biochemical tests: plasma or 24-h urine metanephrines
        ii. Imaging tests: CT or MRI abdomen, Meta-iodobenzylguanidine (MIBG) scan
    d. Treatment: alpha blockade and then adrenalectomy
    e. Monitor for recurrence with urine or plasma metanephrines, and CT or MRI abdomen
3. Additional Manifestations
    a. MEN Type 2A
        i. Primary Hyperparathyroidism
            a.) Manifests in 20–30% of these patients
            b.) May be due to single adenoma vs. multigland hyperplasia
            c.) Diagnosis: elevated serum calcium and parathyroid hormone levels
            d.) Surgical Treatment: parathyroidectomy (extent may vary)
            e.) Monitor with calcium and parathyroid hormone levels
        ii. Cutaneous lichen amyloidosis
        iii. Hirschsprung's disease
    b. MEN Type 2B
        1. Mucosal neuromas
        2. Marfanoid habitus

C. Diagnosis
1. De novo presentation may be medullary thyroid cancer or pheochromocytoma
2. If genetic history is known
    a. Genetic testing for Ret mutation of various codons—some are considered more aggressive than others
    b. Prophylactic total thyroidectomy with central neck dissection by age 1–5
        i. By age 1 for level 3 codon mutations-most aggressive, often will have microscopic tumor already by age 1
        ii. By age 5 for level 2 or level 1 codon mutations
    c. Monitor calcitonin, CEA, and neck ultrasounds yearly
    d. Start screening for pheochromocytoma by age 5 with yearly metanephrines and abdominal imaging

## Additional Notes

The hereditary nature of Multiple Endocrine Neoplasia syndromes makes it important to consider other family members when treating a child with MEN syndrome. Often parents will be aware that the condition runs in their family. After a child has been diagnosed with MEN, it is important to screen siblings. Consultation with a geneticist will help with preparing for the implications of a positive finding on genetic screening. For example, diagnosis of MEN2A indicates that prophylactic total thyroidectomy is necessary by age 5-years-old. Operation can carry complications such as recurrent laryngeal nerve dissection, or hypocalcemia [1]. A longitudinal study of a large proband with MEN2A syndrome has demonstrated that prophylactic total thyroidectomy leads to long-term protection against thyroid cancer [2].

Treating children with MEN syndrome may reveal some features that are different from the disease in adults. A recent report stated that the marfanoid stature present in adults with MEN2B was not present in a series of children with MEN2B, who actually had short stature [3].

## Study Questions

1) The most common pancreatic neuroendocrine tumor in MEN Type 1 patients is:

   a. Insulinoma
   b. Gastrinoma
   c. Non-functioning tumor
   d. Glucagonoma

Answer to Question 1: (c.) Non-functioning tumor. The most common pancreatic neuroendocrine tumor in MEN Type 1 syndrome is a non-functioning tumor. The most common functioning pancreatic neuroendocrine tumor in MEN Type 1 is a gastrinoma.

2) For patients with aggressive mutations of RET in MEN Type 2 syndromes, total thyroidectomy should be performed by age:

   a. 5
   b. 1
   c. 10
   d. 2

Answer to Question 2: (b.) 1. For patients with level 1 or level 2 mutations, which are lower risk than level 3, they should undergo thyroidectomy by age 5. For patients with level 3 codon mutation, which are the most aggressive, they should undergo thyroidectomy by age 1.

3) Which of the following are correct in terms of most common pathology within each MEN syndrome?
   a. Medullary thyroid carcinoma/MEN Type 2A
   b. Gastrinoma/MEN Type 1
   c. Pheochromocytoma/MEN Type 2B
   d. Prolactinoma/MEN Type 1

Answer to Question 3: (a.) Medullary thyroid carcinoma/MEN Type 2A. Primary hyperparathyroidism is the most common manifestation of MEN Type 1, with nearly 90% of patients developing primary hyperparathyroidism by age 40 and nearly 100% by age 50. Medullary thyroid carcinoma is the most common manifestation of MEN Type 2A and 2B, with nearly 90% of patients developing medullary thyroid carcinoma. Pheochromocytoma is also present in MEN Type 2A and 2B, although only 50% of patients will develop it.

## References

1. Tartik A, Arkan G, Kaya C, Karabulut R, Turkyilmaz Z, Sonmez K. Prophylactic thyroidectomy in multiple endocrine neoplasia type 2 syndrome. Gazi Med J. 2021;32(4):589–90.
2. Grubbs EG, Lechan RM, Edeiken-Monroe B, Cote GJ, Trotter C, Tischler AS, Gagel RF. Hereditary endocrine tumours: current state-of-the-art and research opportunities: early thyroidectomy in multiple endocrine neoplasia: a four decade experience. Endocr Relat Cancer. 2020;27(8):T1–8.
3. van den Broek MFM, van Santen HM, Valk GD, Verrijn Stuart AA. Children with multiple endocrine neoplasia type 2B: not tall and marfanoid, but short with normal body proportions. Clin Endocrinol. 2021;95(3):453–9.

## *Further Reading*

Brandi ML, Gagel RF, Bilezikian JP, et al. CONSENSUS: guidelines for diagnosis and therapy of MEN type 1 and type 2. J Clin Endocrinol Metabol. 2001;86(12):5658–71.

Carney JA. Familial multiple endocrine neoplasia: the first 100 years. Am J Surg Pathol. 2005;29(2):254–74.

Jensen RT, Ito T. Gastrinoma. [Updated 2020 Nov 21]. In: Feingold KR, Anawalt B, Boyce A, et al., editors. Endotext. South Dartmouth, MA: MDText.com. 2000. https://www.ncbi.nlm.nih.gov/books/NBK279075/. Accessed 22 Nov 2021.

# Thyroid Nodules

**Robert M. Tuliszewski Jr and Aileen P. Wertz**

**Key Points**

1) Pediatric thyroid nodules are often asymptomatic. Nodules that are found to be malignant have with a higher likelihood of locoregional metastasis to the lateral neck or distant metastasis at presentation when compared to adults.
2) US-guided fine needle aspiration (FNA) is recommended for all thyroid nodules (regardless of size) with concerning features including hypoechogenicity, increased vascularity, irregular margins, microcalcifications, or enlarged regional lymph nodes, or any nodule without concerning features if 1 cm or enlarging on serial ultrasounds.
3) Treatment for malignant nodules includes total thyroidectomy, with thyroid lobectomy reserved for indeterminate nodules, benign nodules, and autonomous nodules that meet specific criteria.

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1. Epidemiology
    a. Incidence:
        i. Thyroid nodules occur in 1–2% of children and increase up to 13% in older adolescents and young adults, with a female to male preponderance (5:1) in this subgroup [1–3]. Though nodules are less common in children than adults, they are more likely to be malignant (22–33%) [4, 5]. Thyroid cancer represents 1–1.5% of all pediatric malignancies with the incidence increasing from 1% in 0–14-year-old children to 7% in 15–19-year-old adolescents [6–8]. Most thyroid cancers are well differentiated with papillary thyroid cancer (PTC) occurring in approximately 85–90% of cases. Follicular thyroid carcinoma (FTC) is uncommon, while medullary thyroid carcinoma (MTC) is a rare variant, and poorly differentiated/anaplastic thyroid carcinoma is truly unusual [9, 10].
    b. Etiology:
        i. Several risk factors are associated with development of thyroid nodules in children, including iodine deficiency, prior radiation exposure, history of thyroid disease, and several genetic syndromes. The major risk factor for PTC is radiation exposure to the thyroid, often from treatment for separate head and neck malignancy one to two decades prior to PTC diagnosis. Iodine deficiency is a clear risk factor for FTC. MTC is often inherited as a dominant mutation in the *RET* proto-oncogene in MEN2A, MEN2B, and familial MTC. An estimated 5% of patients have a family history of nonmedullary thyroid cancer [3].
2. Pathophysiology:
    a. Anatomy:
        i. The thyroid is a bilobed gland with each lobe located lateral to the upper trachea and inferior larynx. The lobes are joined by an isthmus that lies over tracheal rings 2–4. In some patients there is a superior extension to the isthmus called the pyramidal lobe.
        ii. The superior laryngeal nerve (SLN) divides into internal and external branches with the more superior internal branch providing laryngeal sensation and the more inferior external branch in close proximity to the superior thyroid artery. The external branch of the SLN provides motor input to the cricothyroid muscle, located medial to the superior pole of each thyroid lobe.
        iii. The recurrent laryngeal nerve provides motor innervation to the intrinsic laryngeal muscles and is 1–2 cm lateral to the trachea inferiorly. It may pass anterior or posterior to the inferior thyroid artery as it ascends to enter the larynx at the cricothyroid juncture. The RLN may be nonrecurrent approximately 1% of the time and be found more superiorly near the superior thyroid vessels.

- iv. The arterial supply of the thyroid consists of the superior thyroid artery, a branch of external carotid artery, that enters the superior pole of the gland, and the inferior thyroid artery, a branch of the thyrocervical trunk, that enters the gland inferior to the cricothyroid juncture. The inferior thyroid artery supplies both the superior and inferior parathyroid glands. Superior and middle thyroid veins drain into the internal jugular vein and the inferior thyroid vein drains into the innominate vein.
- v. The lymphatic drainage of the superior thyroid is into the superior pretracheal and cervical lymph nodes, and the drainage of the inferior thyroid is into the paratracheal and lower deep cervical nodes.
- vi. The superior parathyroid glands are often located on the posterior aspect of the thyroid near the cricothyroid joint. The inferior parathyroid glands are more variable in location, but are often adjacent to the inferior pole of the thyroid.

3. Clinical features:

   a. In cases of malignant thyroid nodules, children typically present with advanced disease, with regional lymph node involvement occurring in 70% of cases, and distant metastasis to lungs or bones present in up to 20% of cases. In addition, children have higher rates of local and distant recurrence. Despite the often-advanced presentation, prognosis is excellent with long-term survival >90% [11, 12].

4. Diagnosis

   a. Symptoms:

      i. The vast majority of thyroid nodules are asymptomatic and are often detected incidentally by a parent or physician during a routine physical examination. Many patients present with cervical adenopathy as the initial symptom, if malignancy is present. Patients with fixed nodules, cervical adenopathy, or vocal cord paralysis are at high risk for malignancy.

   b. Examination:

      i. A detailed patient history including symptoms of hyper- or hypothyroidism, history of radiation, and family history is required. Physical exam should include visual inspection and palpation of the thyroid gland, palpation of the anterior and lateral neck for adenopathy, and voice assessment. If there is concern for potential vocal cord paralysis, a laryngeal exam should be performed.

   c. Laboratory Tests:

      i. TSH and reflexive T4 should initially be obtained in all patients to determine the functional status of the nodule. A suppressed TSH is indicative of a “hot nodule” or hyperfunctioning nodule that may be indicative of Graves’ disease.

    ii. If there is suspicion for an autoimmune condition such as Hashimoto's thyroiditis or Graves' disease, antithyroid peroxidase antibody testing should be performed.
    iii. If there is suspicion for medullary thyroid carcinoma due to a positive family history, calcitonin and CEA levels should be obtained. For patients at risk for multiple endocrine neoplasia (MEN), genetic testing should be completed at birth for those at risk for MENIIb and by 1 year of age at risk for MENIIa [13, 14].

  d. Radiographic Studies:

    i. Thyroid ultrasound (US) is indicated for palpable nodules, thyroid asymmetry, and/or abnormal cervical adenopathy.
    ii. Concerning features on US include hypoechogenicity, increased vascularity, irregular margins, microcalcifications, size > 4 cm, and enlarged regional lymph nodes.
    iii. CT and/or MRI with contrast is not routinely recommended but can be considered for large or fixed thyroid masses, rapidly expanding tumors, extrathyroidal disease, vocal cord paralysis, or bulky metastatic lymphadenopathy.

5. Treatment

  a. Initial Measures:

    i. US-guided fine needle aspiration (FNA) is indicated for any nodule with suspicious features, a nodule that is >1 cm or shows enlargement on serial US [15].
    ii. Inadequate or non-diagnostic FNA should be repeated with US guidance in 3–6 months.
    iii. Benign nodules should be followed with serial US in 6–12 months and then yearly. A FNA should be undertaken for any observed growth or new suspicious findings.

  b. Subsequent Measures:

    i. In the setting of indeterminate or suspicious findings on FNA, a comprehensive neck US to interrogate the central neck and bilateral lateral neck should be completed to evaluate for regional metastasis. Any enlarged or concerning lymph nodes should subsequently undergo FNA with thyroglobulin washout to aid in identification of metastasis.

  c. Preoperative Preparation:

    i. Prior to thyroidectomy, preoperative laryngeal examination via awake flexible laryngoscopy to assess vocal fold motion is recommended.
    ii. The use of intraoperative laryngeal nerve monitoring is recommended for patients <10 years of age, those undergoing central neck dissection, and repeat surgical procedures, however many surgeons prefer to use monitoring on all thyroid cases.

d. Operative Treatment:

   i. PTC—Total thyroidectomy is recommended as PTC is often multifocal and bilateral. A central neck dissection is indicated if there is clinical evidence of gross extrathyroidal extension and/or locoregional metastasis, and a comprehensive lateral neck dissection is indicated for patients with cytological evidence of metastasis.
   ii. FTC—A thyroid lobectomy is recommended as initial surgical option as FTC is more often unifocal and rarely demonstrates regional metastasis. This should be converted to a total thyroidectomy if there is clear evidence of vascular invasion, extracapsular invasion, tumor size > 4 cm, or distant metastasis.
   iii. MTC—Total thyroidectomy is recommended as early as possible prior to local and distant metastasis. In the setting of MENIIb, prophylactic total thyroidectomy with central neck dissection is recommended as early as possible. A prophylactic total thyroidectomy should be offered before 5 years of age in the setting of MENIIa [16, 17]. In patients with a positive RET mutation a prophylactic total thyroidectomy may be offered prior to development of clinically apparent disease versus close surveillance.
   iv. Indeterminate Nodules—For FNA results that report as atypia or follicular lesion of undetermined significance (AUS/FLUS) or suspicious for follicular/Hurthle neoplasm, recommendation is for thyroid lobectomy plus isthmusectomy as this cohort has been found to have higher rates of malignancy versus adults with similar FNA results.
   v. Benign nodules—Thyroid lobectomy may be considered due to increasing size, compressive symptoms, cosmetic reasons, or parent/child choice.
   vi. Autonomous nodules—For patients with suppressed TSH and confirmed uptake on thyroid scintigraphy of a “hot nodule,” thyroid lobectomy is recommended.

e. Post-operative Treatment:

   i. Thyroid hormone replacement must be prescribed in the absence of functional thyroid tissue.
   ii. Post-operative calcium and PTH levels should be obtained to monitor for any signs of hypocalcemia and/or hypoparathyroidism. Many patients are placed on routine calcium and vitamin D supplementation prophylactically.
   iii. Patients with PTC should be stratified into low-, intermediate-, and high-risk categories based on surgical pathology. Patients with disease confined to thyroid or with incidental microscopic metastasis are classified as low risk. Patients with central neck or limited lateral metastasis are classified as intermediate risk, and patients with extrathyroidal invasion, extensive regional metastasis, with or without distant metastasis are classified as high risk.

iv. Low-risk patients should undergo post-operative staging with a TSH suppressed thyroglobulin testing within 12 weeks after surgery. Initial TSH goal for these patients is 0.5–1.0 mIU/l.

v. Intermediate-risk and high-risk patients should undergo a TSH-stimulated thyroglobulin level and diagnostic $^{123}$I whole body scan within 12 weeks after surgery. This will help to determine the presence of residual disease and need for radioactive $^{131}$I treatment. Initial TSH goals include 0.1–0.5 mIU/l for intermediate-risk and <0.1 mIU/l for high-risk patients.

vi. Radioactive $^{131}$I treatment is indicated for treatment of iodine-avid persistent locoregional or nodal disease that cannot be resected, invasion of vital structures, and for iodine-avid distant metastases. $^{131}$I treatment is typically administered 4–6 weeks post-operatively.

vii. To facilitate $^{131}$I uptake in iodine-avid disease, TSH should be >30 mIU/l. To achieve this level, recommendation is to hold levothyroxine for >14 days.

viii. Long-term surveillance is recommended with US obtained 6 months post-operatively and then annually for 5 years for low-risk patients and every 6–12 months for intermediate- and high-risk patients. Any suspicious or changing nodules on US should undergo FNA as previously described.

ix. Thyroglobulin is a sensitive tumor marker for patients with differentiated thyroid cancer as it is synthesized by thyroid follicular cells and should be undetectable after complete thyroidectomy. Thyroglobulin antibodies should be measured simultaneously as the presence of thyroglobulin antibodies will interfere with thyroglobulin measurements, with thyroglobulin antibodies present in approximately 25% of patients. If thyroglobulin antibodies are detected, they can be used as a tumor marker similar to thyroglobulin. An undetectable thyroglobulin level or level < 0.1 ng/ml is indicative of disease remission. A rising thyroglobulin level or value between 0.1 and 10 ng/ml should prompt further workup with US. A level > 10 ng/ml may be indicative of recurrence and requires imaging with US and CT neck and chest. Thyroglobulin levels should be obtained every 3–6 months for 2 years in low-risk patients and for 3 years in intermediate- and high-risk patients. Thyroglobulin should be obtained annually thereafter.

x. The majority of disease recurrence in children is in cervical lymph nodes. For macroscopic disease recurrence >1 cm surgical resection is preferable to $^{131}$I treatment. For recurrence <1 cm $^{131}$I treatment, observation with TSH suppression, or surgical resection are all potential options.

xi. For patients with FTC long-term surveillance includes monitoring thyroglobulin levels, physical exam, and serial neck US's similar to recommendations for PTC.

   xii. In the setting of MTC, long-term surveillance should include calcitonin and CEA levels as well as serial neck US and body imaging with chest X-ray and CT scans of neck and chest. RAI does not have a role in post-operative treatment of MTC as the malignant parafollicular C cells do not take up iodine.

f. Complications:

   i. The most common complication after thyroidectomy is transient or permanent hypocalcemia occurring in 5–15% of patients [18]. The most feared complication is RLN injury which occurs in approximately 1–6% of cases [19, 20]. In patients undergoing thyroid lobectomy, hypothyroidism can be a long-term sequela in 6.5–49% [21].
   ii. Patients <10 years of age have a higher risk of complications associated with extrathyroidal extension, lymph node dissection, and repeat surgery [19, 22, 23].

## Additional Notes

Though a rare type of pediatric cancer overall, the incidence of thyroid cancer in children is increasing, particularly in adolescents. In addition, thyroid nodules in children have a higher likelihood of being malignant when compared to adults. And when thyroid cancer is detected, pediatric patients have a higher likelihood of presenting with a larger tumor volume in the thyroid gland itself, as well as increased likelihood of having locoregional metastasis to the central or lateral neck or distant metastasis to the lungs. Combining this with increased and more sensitive and specific genetic testing for mutations that are high risk for thyroid cancer, early treatment is recommended for thyroid nodules that meet specific criteria, patients at risk for future malignancy, and diagnosed thyroid cancer. By following such guidelines, the prognosis for thyroid malignancy is quite good with long-term survival greater than 90% [13].

As the knowledge of thyroid nodules and malignancy continues to expand it is imperative that treatment is coordinated through centers experienced in the workup, treatment, and post-operative surveillance. Pediatric patients have a higher local and distant recurrence rates with the recurrence-free 20-year survival rate is 10.1% for patients younger than 10 years, and 48.3% in patients older than 10 years [24]. Such data necessitates long-term follow-up with appropriate testing. As recommendations have moved toward aggressive surgical treatment for varying pathology, it is important to note that there are lower general complication rates (8.7 vs. 13.4%) and endocrine complication rates (5.6 vs. 11%) by high volume endocrine surgeons [25].

The role of post-operative RAI for thyroid cancer has been a debate in the pediatric population with previous recommendations advocating for RAI in all patients as this has been correlated with decreased rates of thyroid bed and lymph node recurrence. The long-term sequela of RAI treatment including xerostomia, pulmonary fibrosis, menstruation abnormalities, decreased sperm count, and secondary cancers has pushed to establish risk categories for patients with thyroid cancer to determine which patients require post-operative RAI. Current recommendations include RAI for distant metastasis, extensive regional lymph node involvement, and unresectable disease.

## Study Questions

1) In children mutations in _____ are most common in medullary thyroid carcinoma, while mutations in _____ are most common in papillary thyroid carcinoma.

   a. RET, BRAF
   b. RAS, BRAF
   c. BRAF, RAS
   d. RET, RET

   Answer to Question 1: (d.) RET, RET

   Mutations in the RET proto-oncogene are responsible for three inheritable syndromes associated with MTC: MEN-2A has RET mutations in exons 10 and 11, while mutations in exon 16 lead to MEN-2B. Familial MTC is though to be a less severe variant of MEN-2A. While in adults a mutation in BRAF is most associated with PTC, in children a mutation in RET is most commonly observed in PTC.

2) A 16-year-old female presents to your clinic for follow-up of an incidental right thyroid nodule identified on CT imaging obtained after a MVC. She is asymptomatic and physical exam does not reveal any palpable thyroid abnormality or neck adenopathy. An US-guided FNA is obtained which reveals a 0.7 cm hypoechoic nodule in the inferior thyroid lobe. FNA is reported as atypia of undetermined significance. The next step in treatment is:

   a. Perform genetic testing on FNA sample
   b. Repeat FNA in 3–6 months
   c. Thyroid lobectomy plus isthmusectomy
   d. Observe and repeat imaging in 6 months

   Answer to Question 2: (c.) Thyroid lobectomy plus isthusectomy

   ATA guidelines recommend definitive surgery for indeterminate FNA findings in children due to the increased risk of malignancy of these results (approximately 35%) compared to adults (5–15%). Due to limited data, genetic testing on FNA results has not been verified in children as it has been in adults. Repeat FNA in 3 months is the recommendation for FNA results consistent with insufficient or nondiagnostic sample.

3) A 7-year-old male presents to your clinic with complaints of dry eyes and thyroid enlargement. Recent lab values in his chart show a depressed TSH and elevated T4. You recommend an endocrine evaluation and neck US which reveal a 1.0 cm right mid pole thyroid nodule. Further workup is positive for thyroid peroxidase antibodies and scintigraphy reveals a "hot" nodule. The next step of treatment includes:

   a. Methimazole managed by his endocrinologist
   b. FNA of the nodule
   c. Repeat US in 6 months to monitor any change in size of thyroid nodule
   d. Thyroid lobectomy

Answer to Question 3: (d.) Thyroid lobectomy

In pediatric patients with suppressed TSH, an associated "hot" thyroid nodule on thyroid scintigraphy, thyroid lobectomy is the recommended treatment. In children up to 1/3 of autonomous thyroid nodules may contain a differentiated thyroid cancer. A FNA can be considered if the nodule has concerning features. Observation can be offered for asymptomatic patients and subclinical hyperthyroidism.

## References

1. Niedziela M, et al. A prospective study of thyroid nodular disease in children and adolescents in Western Poland from 1996 to 2000 and the incidence of thyroid carcinoma relative to iodine deficiency and the Chernobyl disaster. Pediatr Blood Cancer. 2004;42(1):84–92. https://doi.org/10.1002/pbc.10421.
2. Oertel JE, Klinck GH. Structural changes in the thyroid glands of healthy young men. Med Ann Dist Columbia. 1965;34:75–7.
3. Francis GL, et al. Management guidelines for children with thyroid nodules and differentiated thyroid cancer. Thyroid. 2015;25(7):716–59. https://doi.org/10.1089/thy.2014.0460.
4. Gupta A, et al. A standardized assessment of thyroid nodules in children confirms higher cancer prevalence than in adults. J Clin Endocrinol Metab. 2013;98(8):3238–45. https://doi.org/10.1210/jc.2013-1796.
5. Niedziela M. Pathogenesis, diagnosis and management of thyroid nodules in children. Endocr Relat Cancer. 2006;13(2):427–53. https://doi.org/10.1677/erc.1.00882.
6. Garcia CJ, et al. Sonography of multinodular thyroid gland in children and adolescents. Am J Dis Child. 1992;146(7):811–6. https://doi.org/10.1001/archpedi.1992.02160190043017.
7. Khurana KK, et al. The role of fine-needle aspiration biopsy in the management of thyroid nodules in children, adolescents, and young adults: a multi-institutional study. Thyroid. 1999;9(4):383–6. https://doi.org/10.1089/thy.1999.9.383.
8. Lugo-Vicente H, et al. Pediatric thyroid nodules: management in the era of fine needle aspiration. J Pediatr Surg. 1998;33(8):1302–5. https://doi.org/10.1016/s0022-3468(98)90174-9.
9. Jatana KR, Zimmerman D. Pediatric thyroid nodules and malignancy. Otolaryngol Clin North Am. 2015;48(1):47–58.
10. Roy R, Kouniavsky G, Schneider E, et al. Predictive factors of malignancy in pediatric thyroid nodules. Surgery. 2011;150(6):1228–33.
11. Yoskovitch A, Laberge JM, Rodd C, et al. Cystic thyroid lesions in children. J Pediatr Surg. 1998;33(6):866–70.

12. Robie DK, Dinauer CW, et al. The impact of initial surgical management on outcome in young patients with differentiated thyroid cancer. J Pediatr Surg. 1998;33(7):1134–8.
13. Skinner MA, Wells SA. Medullary carcinoma of the thyroid gland and the MEN 2 syndromes. Semin Pediatr Surg. 1997;6:134.
14. Telander RL, Moir CR. Medullary thyroid carcinoma in children. Semin Pediatr Surg. 1994;3:188.
15. Iakovou I, Giannoula E, Sachpekidis C. Imaging and imaging-based management of pediatric thyroid nodules. J Clin Med. 2020;9(2):384.
16. Lallier M, St-Vil D, et al. Prophylactic thyroidectomy for medullary thyroid carcinoma in gene carriers of MEN2 syndrome. J Pediatr Surg. 1998;33:846.
17. La Quaglia MD, Telander RL. Differentiated and medullary thyroid cancer in childhood and adolescence. Semin Pediatr Surg. 1997;6:42.
18. Guille JT, Opoku-Boateng A, et al. Evaluation and management of the pediatric thyroid nodule. Oncologist. 2015;20(1):19–27.
19. Sosa JA, Tuggle CT, et al. Clinical and economical outcomes of thyroid and parathyroid surgery in children. J Clin Endocrinol Metab. 2008;93:3058–65.
20. Kundel A, Thompson GD, et al. Pediatric endocrine surgery: a 20 year experience at the Mayo Clinic. J Clin Endocrinol Metab. 2014;99:399–406.
21. Bryarly RC, Shockley WW, Stucker FJ. The method of management of thyroid surgery in the pediatric patient. Laryngoscope. 1985;95:1025.
22. Demidchik YE, Demidchik EP, et al. Comprehensive clinical assessment of 740 cases of surgically treated thyroid cancer in children of Belarus. Ann Surg. 2006;243:525–32.
23. Bargren AE, Meyer-Rochow GY, et al. Outcomes of surgically managed pediatric thyroid cancer. J Surg Res. 2009;156:70–3.
24. Alessandri AJ, Goddard KJ, et al. Age is the major determinant of recurrence in pediatric differentiated thyroid carcinoma. Med Pediatr Oncol. 2000;35:41.
25. Tuggle CT, Roman SA, et al. Pediatric endocrine surgery: who is operating on our children? Surgery. 2008;144:869–77.

## *Further Reading*

Francis GL, et al. Management guidelines for children with thyroid nodules and differentiated thyroid cancer. Thyroid. 2015;25(7):716–59. https://doi.org/10.1089/thy.2014.0460.

Gupta A, et al. A standardized assessment of thyroid nodules in children confirms higher cancer prevalence than in adults. J Clin Endocrinol Metab. 2013;98(8):3238–45. https://doi.org/10.1210/jc.2013-1796.

Niedziela M. Pathogenesis, diagnosis and management of thyroid nodules in children. Endocr Relat Cancer. 2006;13(2):427–53. https://doi.org/10.1677/erc.1.00882.

# Breast Lesions in Adolescents

## Rebecca Michelle Jordan and Nicole E. Sharp

**Key Points**
1. Understand normal and abnormal development of the breast in children and adolescents.
2. Recognize clinical and diagnostic findings associated with common breast lesions in adolescents.
3. Understand the diagnosis and treatment algorithm for common breast lesions.
4. Be familiar with the common surgical techniques utilized for breast lesions.

1. Background

    a. Breast development begins between gestational weeks four and six with formation of ectodermal ridges at “Hughes line” or the milk line spanning from the axilla to groin. These ridges recede over the next few weeks except those at the fourth anterior intercostal space with proliferation of the primary mammary bud beginning at week 10. After birth, the primary breast bud will involute due to withdrawal of maternal estrogen.
    b. Thelarche marks the beginning of puberty with proliferation of ducts and fat stimulated by estrogen between ages 8 and 13 years. Timing and coordinated development of each breast bud varies by individual and race.

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   c. Supernumerary nipples or accessory breast tissue can develop anywhere along the milk line, with accessory tissue most common in the axilla. Asymmetry is common in adolescents with a development differential of up to 2 years between breasts. Hypoplasia or aplasia may occur with Poland syndrome which affects chest wall and upper extremity development while atrophy may develop due to other underlying causes such as scleroderma, malnutrition, eating disorders, endocrine disorders, or trauma and incisions to the infant breast bud. Adolescent hypertrophy presents with excessive stromal hyperplasia during breast development causing severe macromastia which can lead to back and neck strain. Surgical reduction may be considered in older adolescents.

2. Workup

   a. While breast masses in adolescents are rare and frequently benign, it is important to include uncommon, high-risk, and malignant lesions within the differential diagnosis. All lesions that present in adults can present in the adolescent breast, including atypical ductal hyperplasia, atypical lobular hyperplasia, papillary lesions, radial scars, spindle cell lesions, mucoceles, adenomas, papillomas, and pseudoangiomatous stromal hyperplasia (PASH). Common dermatologic lesions such as eczema, psoriasis, and acne are also frequently seen in the adolescent population and can cause significant distress when presenting on the breast.
   b. During the clinical breast exam, begin by visually evaluating bilateral breasts in upright and supine positions looking for any asymmetry. Bilateral breast should be palpated completely noting tissue density changes or palpable masses. Evaluate for overlying skin changes, erythema, nipple discharge, and tenderness. Palpate bilateral axillae for accessory tissue or lymphadenopathy. Location of an identified lesion is described using the clockface method about the nipple.
   c. Ultrasound is the preferred imaging modality when evaluating a suspicious breast mass in adolescents. Mammogram is infrequently utilized in the pediatric populations due to breast density and radiation exposure.
   d. Image-guided percutaneous core needle biopsy is recommended as an initial diagnostic procedure for breast lesions. Pathology should be evaluated for radiographic and clinical concordance. If there is concern for discordant findings, further histological evaluation should be considered with repeat core needle biopsy, vacuum biopsy, or excisional biopsy. The decision to pursue surgical excision is often a shared decision with the surgeon, patient and their family based on clinical, radiographic, and pathologic findings along with the patients' risk factors and anxiety.

3. Adolescent Breast Lesions

   a. Fibroepithelial lesions

      i. Epidemiology

1. Fibroepithelial lesions are composed of both fibroadenomas and phyllodes tumors. Fibroadenomas are the most common benign breast mass in adolescents comprising up to 95% of surgically removed pediatric breast masses. These most commonly occur after age 14 with increased prevalence from 15 to 16 years of age but may present at earlier ages in African American females. Phyllodes tumors are most common in middle-aged women but have been reported in children as young as 10 years old.

ii. Pathophysiology

1. Fibroepithelial lesions result from the abnormal growth of fibrous and glandular tissue in the breast. Juvenile fibroadenomas occur in children and adolescents near puberty while adult fibroadenomas can also affect adolescents and young adults. In comparison, juvenile fibroadenomas tend to be larger in size. Pathophysiology is unclear but presumed due to an exaggerated response to estrogen.
2. Phyllodes tumors are stromal tumors of the breast and may be benign, borderline, or malignant. While most are histologically benign, phyllodes tumors are the most common malignant breast mass in adolescents. Histologically they are like fibroadenomas but show more stromal proliferation and have the potential to recur and metastasize.

iii. Clinical Features

1. Fibroadenomas often range in size from 1 to 3 cm and may vary with hormonal fluctuations like menses, birth control, or pregnancy. Up to 25% of patients present with multiple lesions. Giant juvenile fibroadenomas weigh more than 500 g, comprise more than 80% of the breast, or grow to more than 5–10 cm.
2. Phyllodes tumors are difficult to distinguish from giant fibroadenomas but should be suspected if the lesion is rapidly growing or over 5 cm.

iv. Diagnosis

1. Clinical exam and imaging is nearly indistinguishable between a fibroadenoma and phyllodes tumor. Fibroadenomas present as a smooth, mobile, rubbery mass with distinct borders on palpation. Findings of a large, painless mass are concerning for giant fibroadenoma or phyllodes tumor. With larger masses, the overlying skin may be stretched from rapid growth.
2. On ultrasound, fibroadenomas will appear as a round, oval, or macrolobulated mass with circumscribed margins and uniform hypoechogenicity. Phyllodes tumors may include lobulations and heterogenous sonographic pattern and often lacks microcalcifications.
3. For concerning lesions, ultrasound-guided core needle biopsy can be performed to distinguish phyllodes tumor from fibroadenoma.

v. Treatment

1. Findings on history and physical consistent with an asymptomatic fibroadenoma that is less than 3 cm may be monitored with serial ultrasounds at 6 months, 1 year, and 2 years. Surgical excision should be considered for lesions where there is inability to rule out a phyllodes tumor particularly in those lesions over 5 cm or with enlargement. Excision may also be considered for those patients with symptomatic or cosmetic concerns. Consideration should be given to cosmetic placement of incisions just above the inframammary fold, periareolar, or axilla. Dissection should be carried directly down to the lesion and care should be taken to avoid excision of surrounding breast parenchyma to avoid resultant asymmetry. Most fibroadenomas, even those of substantial size, can be shelled out with excellent cosmetic results as the surrounding breast tissue will fill in the defect. Counseling of the patient and their families must include potential damage to the breast bud and resultant asymmetry.
2. No margins are needed for fibroadenomas or benign phyllodes tumors. Margins for borderline and malignant phyllodes are at least 1 cm per National Comprehensive Cancer Network version 3. 2022. There is no indication for axillary staging. Breast conservation preferred whenever possible. Close margins are associated with increased recurrence risk but are not an absolute indication for mastectomy.

b. Fibrocystic change

i. Epidemiology

1. Fibrocystic change is present in up to 50% of adolescent females.

ii. Pathophysiology

1. The etiology of fibrocystic change is unclear but thought to be secondary to an imbalance in estrogen and progesterone, primarily throughout the menstrual cycle.

iii. Clinical features

1. Clinical presentation ranges from asymptomatic to painful breast tissue before menses with improvement during menses. Most clinical complaints are due to breast thickening or lump.

iv. Diagnosis

1. Palpable tissue findings may be vague or ridge-like areas of density or thickening. Non-discrete lumps may be noted, most often in the upper outer quadrants. Cordlike nodularity with associated cysts can also be palpated which will collapse each month with menses.
2. Fibrocystic change is a clinical diagnosis based on history and physical exam and lack of imaging findings consistent with other diagnoses.

v. Treatment

1. Primary treatment for fibrocystic change is reassurance. If symptoms persist, analgesia with non-steroidal anti-inflammatory medications or oral contraceptives can be trialed. Additional lifestyle modifications should be considered such as reduction of caffeine intake and wearing a well-fitting bra.

c. Cysts

i. Epidemiology

1. Benign cysts are common in adolescents.

ii. Pathophysiology

1. A cyst is a fluid filled cavity lined by epithelium within surrounding breast tissue that may be secondary to duct obstruction or dilation of lobular acini. Retroareolar cysts occur with obstruction of the Montgomery areolar tubercle, a sebaceous gland associated with the terminal portion of a lactiferous duct. Morgagni tubercles are periareolar glands that may intermittently drain clear to brown fluid but may also present as a subareolar mass.

iii. Clinical features

1. Since adolescents do not undergo screening exams, these are most often found on physical exam but may also be seen incidentally on imaging exams performed for other reasons. Symptoms may range from asymptomatic to a palpable mass which can fluctuate in size with menstruation. Retroareolar cysts may be asymptomatic or have associated inflammatory changes.

iv. Diagnosis

1. Patients may or may not have a palpable breast mass or other findings on physical exam.
2. Ultrasound should be performed for a palpable mass. Simple cysts will be round, well-circumscribed, and anechoic. Complex cysts may have thick walls, intracystic masses, or discrete solid components.

v. Treatment

1. Simple cysts that are asymptomatic may be observed. Follow-up ultrasound can be performed following menstruation to ensure resolution and stability.
2. For persistent, symptomatic, or complex cysts, ultrasound-guided aspiration may be considered. If aspiration reveals clear fluid, this may be discarded. Aspirated fluid which is bloody should be sent for cytology. If purulent fluid is found, this should be treated as an abscess

with antibiotics and surveillance. Any aspirated cyst should be re-evaluated by ultrasound in 3 months to ensure resolution.

3. Surgical excision may be performed rarely for persistent cysts that remain symptomatic or do not resolve with serial aspiration.

d. Inflammatory/Infectious lesions
    i. Epidemiology
        1. Mastitis and abscesses are infectious lesions that present with inflammatory changes to the breast. These have a bimodal distribution seen in children under 2 months and then 8–17 years old.
    ii. Pathophysiology
        1. Pediatric mastitis and abscess most frequently occur due to associated skin infections, instrumentation, or lactation. Other risk factors include tobacco use, piercings, and shaving. The most common bacterial pathogen is Staphylococcus aureus, identified in up to 75% of cases. Other common pathogens include Gram-negative bacilli, Group A Streptococcus, and Enterococcus species.
        2. Fat necrosis is a benign inflammatory entity due to breast trauma that can mimic carcinoma.
    iii. Clinical Features
        1. Patients may report pain, fever, erythema, warmth, drainage, or nipple discharge with focal breast tenderness. History of blunt or penetrating trauma to the breast should be elucidated and may correlate with new findings concerning for fat necrosis.
    iv. Diagnosis
        1. Swelling, skin erythema, warmth, induration, and focal tenderness may be seen with reactive lymphadenopathy in the ipsilateral axilla. Palpate for fluctuance indicative of an underlying abscess.
        2. Ultrasound can be performed to differentiate mastitis from abscess.
    v. Treatment
        1. Antibiotic therapy is the treatment of choice with focus on Staphylococcus aureus coverage. If an abscess is identified, ultrasound-guided aspiration should be attempted first if appropriate. Care should be taken to avoid damage to the breast bud. Follow-up ultrasound should be considered to ensure resolution after aspiration. Incision and drainage may be necessary if aspiration fails or if the abscess cavity is large or loculated.
        2. Fat necrosis is often self-limiting with supportive treatment.

e. Nipple discharge

  i. Epidemiology

    1. Nipple discharge is rare in children and adolescents. However, up to 80% of females will experience discharge during their reproductive age.

  ii. Pathophysiology

    1. Nipple discharge may be physiologic or pathologic. Physiologic discharge is often bilateral, comes from multiple ducts, does not occur spontaneously, and may be secondary to nipple stimulation. Pathologic discharge is spontaneous and usually unilateral. Milky discharge may be physiologic during pregnancy or post-partum states while galactorrhea is inappropriate lactation unrelated to these. Etiologies include medications such as antihypertensives, H2-blockers, oral-contraceptive pills/hormone replacement, opioids, and psychotropic agents, prolactin secreting tumors, hypothyroidism, or chronic renal disease. A prolactinoma may also present with primary or secondary amenorrhea with prolactin level >100 ng/mL suggestive of diagnosis. Bloody discharge can be due to an intraductal papilloma, duct ectasia, or malignancy. Intraductal papillomas occur with proliferation of cells in the mammary ducts projecting into the lumen. These lesions do not always present with nipple discharge but are the most common cause of bloody nipple discharge when present. Purulent drainage is concerning for mastitis or abscess. Duct ectasia can also present with multicolored/sticky drainage while watery discharge may indicate underlying papilloma or cancer. Nipple discharge of any type in males is considered pathologic and further workup is warranted.

  iii. Clinical Features

    1. Patients may present with unilateral or bilateral nipple discharge that is spontaneous or non-spontaneous, noted from multiple ducts versus a single duct. Findings of unilateral, non-spontaneous, single duct discharge, or increasing frequency is concerning for underlying pathologic process.

  iv. Diagnosis

    1. Discharge may or may not be reproducible on exam.
    2. Diagnosis is based on clinical history and physical exam. Medications and recreational drug use should be explored for causes of nipple discharge. If galactorrhea is present, pregnancy should first be ruled out. Workup for pathologic discharge may include serum prolactin and estradiol concentrations, and ultrasound of the affected breast to iden-

tify any underlying masses. A sample of reproducible discharge may be sent for cytology, however this is not routinely recommended as it rarely changes patient management. If galactorrhea is present with an elevated prolactin level, further imaging of the sella turcica is warranted with an MRI of the brain. Breast imaging, typically starting with an ultrasound, may be considered if there is clinical suspicion for a pathologic cause.

v. Treatment

1. Medications should be stopped if thought to be the underlying etiology. If a mass or other abnormality is seen on imaging, consider core needle biopsy or surgical excision. For persistent discharge due to ductal ectasia, definitive management involves surgical excision of the involved duct.

f. Cancer

i. Epidemiology

1. Breast cancer is extremely rare in adolescents. Metastatic disease to the breast is more common than primary breast cancer in adolescents with primary breast carcinoma accounting for less than 1% of childhood cancers and less than 0.1% of breast cancers.

ii. Pathophysiology

1. Primary malignancies that may present with metastatic disease to the breast include hepatocarcinoma, non-Hodgkin lymphoma, leukemia, rhabdomyosarcoma. Secretory adenocarcinoma (formerly juvenile carcinoma) is the most common primary breast cancer in adolescents and is less aggressive than infiltrating ductal carcinoma.
2. There is increased risk of primary breast cancer in children exposed to radiation, especially those receiving mantle radiation for Hodgkins lymphoma in the pubertal years. Other risk factors include family history of deleterious breast and ovarian cancer associated genetic mutations (such as BRCA 1, BRCA 2, TP53 (Li–Fraumeni syndrome), PTEN (Cowden syndrome), or those treated for other childhood cancers.

iii. Clinical features

1. Compared to adult patient, adolescents are more likely to present with a large mass at diagnosis. However, nipple retraction and discharge appear less commonly.

iv. Diagnosis

1. Malignant lesions typically present as a palpable, firm, poorly circumscribed mass. Evaluation for other suspicious findings should include

skin dimpling, fixation to the chest wall, edema, or ulceration and examine the bilateral axilla for any lymphadenopathy.

2. Ultrasound findings may be variable and non-specific. Malignant lesions typically appear as a hypoechoic mass with irregular margins, inhomogeneous internal echoes, and variable posterior acoustic shadowing. Metastatic lesions are frequently multiple and bilateral. Computed tomography is not frequently used to evaluate breast masses in children but may incidentally identify breast metastasis on imaging for children with other known primaries. Image-guided core needle biopsy is performed to obtain a tissue diagnosis.

v. Treatment

  1. Complete surgical resection is the treatment of choice for primary breast malignancies. Specimen margins should follow the NCCN guidelines based on source of the primary tumor.

## Study Questions

1. A 15-year-old female presents to the clinic with complaints of left breast pain. She also notes a “lump” in her left breast that is painful with menses. She reports onset of menarche at age 12 and a family history of breast cancer in her maternal grandmother at age 70. On physical exam, a smooth, mobile mass with round edges 1 cm in size is palpated in the upper outer quadrant. What is the next step in management for this finding?

   a. Computed tomography of the chest
   b. Magnetic resonance imaging of the chest
   c. Ultrasound of the left breast
   d. Core needle biopsy of the palpable lesion
   e. Excisional biopsy given her family history of breast cancer

Answer (c): Ultrasound of the left breast. The patient is presenting with history and physical exam findings most consistent with a fibroadenoma. The next step in management should be an ultrasound to confirm the diagnosis. CT and MRI are not routinely used in the workup of pediatric breast lesions. Image-guided core needle biopsy is the procedure of choice if a tissue diagnosis is needed. However, biopsy is not required in patients that have clinical exams and imaging concordant with a fibroadenoma. If there are suspicious features of a phyllodes tumor, like rapid growth or size > 5 cm, then image-guided biopsy would be recommended.

2. A 17-year-old female complains of swelling to her bilateral axillae which varies in size each month. She denies associated trauma, recent infections, pain, erythema, or drainage. She reports a history of menarche at age 10 and takes no

medications. On physical exam, a soft mound of tissue is palpated in each axilla without findings of associated mass or overlying skin changes. The most likely diagnosis is:

a. Lymphadenopathy
b. Accessory breast tissue
c. Fibroadenoma
d. Abscess
e. Metastatic cancer

Answer (b): Accessory breast tissue. This is most often evident with fluctuations in hormones at puberty or with pregnancy. Accessory breast tissue or supernumerary nipples can be seen anywhere along the milk line with accessory breast tissue most commonly in the axilla. Accessory breast tissue may be asymptomatic or cause pain, cosmetic changes, or difficulty with arm movement. Treatment for asymptomatic disease is supportive. However, surgical intervention may be considered recommended in the symptomatic adults with liposuction, excision, or a combination of both.

3. A 14-year-old female presents with a right breast mass that has rapidly increased in size over the past 3 months. She has noticed that the overlying skin is stretched and shiny but denies pain, erythema, warmth, or nipple discharge. An ultrasound is performed which shows a 6 cm oval mass with lobulations and a heterogenous sonographic pattern. Image-guided biopsy should be recommended to rule out what lesion?

a. Fibroadenoma
b. Fibrocystic disease
c. Simple cyst
d. Phyllodes tumor
e. Adolescent hypertrophy

Answer (d): Phyllodes tumor. This patient is presenting with an enlarging painless mass with overlying skin stretching concerning for giant fibroadenoma or phyllodes tumor. In lesions that present with symptoms, rapid growth or size > 5 cm, there is clinical suspicion for a phyllodes tumor which should prompt an image-guided biopsy. Ultrasound findings include lobulations with a heterogenous sonographic pattern and size over 5 cm is concerning for phyllodes tumor.

## Images

1. Benign phyllodes tumor 2 × 2.5 × 1.5 cm. On ultrasound the lesion is indistinguishable from a fibroadenoma. Core needle biopsy showed a fibroepithelial lesion. Excisional biopsy was consistent with a benign phyllodes tumor.

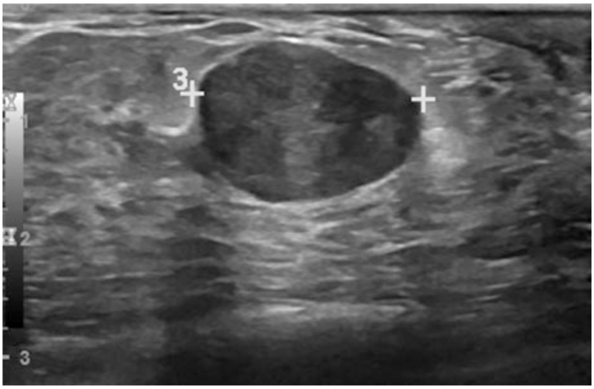

Photo Credit: Geisinger Medical Center Breast Radiology Department, Danville, PA

2. Gross cross section of a fibroadenoma.

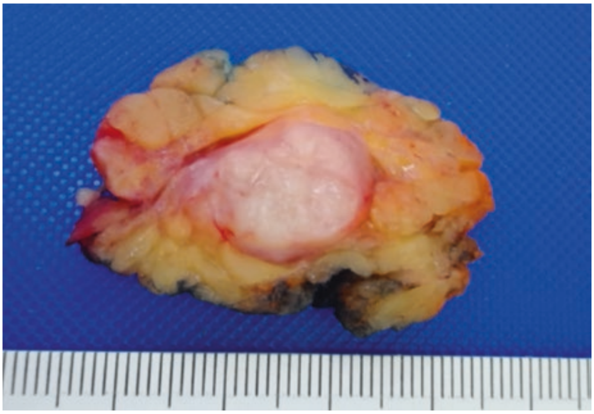

Photo Credit: Geisinger Laboratory Medicine, Surgical Pathology, Danville, PA

## Further Reading

Aydogan F, Baghaki S, Celik V, Kocael A, Gokcal F, Cetinkale O, Unal H. Surgical treatment of axillary accessory breasts. Am Surg. 2010;76(3):270–2.

Brownstone ND, Celie KB, Spigland NA, Otterburn DM. Pediatric breast fibroadenomas: a systematic review and algorithm for treatment. Ann Plast Surg. 2019;83(5):601–5. https://doi.org/10.1097/SAP.0000000000001717.

Chung EM, Cube R, Hall GJ, González C, Stocker JT, Glassman LM. From the archives of the AFIP: breast masses in children and adolescents: radiologic-pathologic correlation. Radiographics. 2009;29(3):907–31. https://doi.org/10.1148/rg.293095010.

Kaneda HJ, Mack J, Kasales CJ, Schetter S. Pediatric and adolescent breast masses: a review of pathophysiology, imaging, diagnosis, and treatment. AJR Am J Roentgenol. 2013;200(2):W204–12.

Kennedy RD, Boughey JC. Management of pediatric and adolescent breast masses. Semin Plast Surg. 2013;27(1):19–22. https://doi.org/10.1055/s-0033-1343991.

Khaja A, DeSilva N. The female adolescent breast: disorders of development. Curr Opin Obstet Gynecol. 2019;31(5):293–7. https://doi.org/10.1097/GCO.0000000000000564.

Klimberg S, Hunt K. Disease of the breast. In: Townsend CM, Beauchamp RD, Evers BM, Mattox K, editors. Sabiston textbook of surgery. 21st ed. Philadelphia, PA: Elsevier Saunders; 2022. p. 809–55.

Nordt CA, DiVasta AD. Gynecomastia in adolescents. Curr Opin Pediatr. 2008;20(4):375–82. https://doi.org/10.1097/MOP.0b013e328306a07c.

Onstad M, Stuckey A. Benign breast disorders. Obstet Gynecol Clin North Am. 2013;40(3):459–73. https://doi.org/10.1016/j.ogc.2013.05.004.

Pandya S, Moore RG. Breast development and anatomy. Clin Obstet Gynecol. 2011;54(1):91–5. https://doi.org/10.1097/GRF.0b013e318207ffe9.

Sabel MS, Helvie MA, Breslin T, Curry A, Diehl KM, Cimmino VM, Chang AE, Newman LA. Is duct excision still necessary for all cases of suspicious nipple discharge? Breast J. 2012;18(2):157–62. https://doi.org/10.1111/j.1524-4741.2011.01207.x.

Templeman C, Hertweck SP. Breast disorders in the pediatric and adolescent patient. Obstet Gynecol Clin North Am. 2000;27(1):19–34. https://doi.org/10.1016/s0889-8545(00)80004-2.

Warren R, Degnim AC. Uncommon benign breast abnormalities in adolescents. Semin Plast Surg. 2013;27(1):26–8. https://doi.org/10.1055/s-0033-1343993.

# Ovarian Masses and Cysts

**Niti Shahi and Muriel A. Cleary**

**Key Points**

1) Ultrasound is the first-line imaging modality for the evaluation of ovarian masses.
2) Size, the Ueland index, and the ovarian crescent sign are important features to distinguish benign from malignant masses on an ultrasound.
3) Ovarian preserving approaches should be utilized, when possible, during surgical resection.
4) Malignant masses require careful staging and excision with oophorectomy, either laparoscopically or open with attention to fertility preservation.

## A. Epidemiology

A. Ovarian tumors in children have a reported incidence of approximately 2.6 per 100,000 [1–4].
B. Only 1% of childhood cancers are ovarian in nature. The majority of ovarian masses are benign tumors with an overall low rate of malignancy ranging from 10 to 20% [5].
C. It has been historically estimated that only 1.2% of all ovarian cancer diagnoses occur in children under the age of 19 [6].

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B. Pathophysiology and Types of Masses: Ovarian masses are categorized into solid or cystic masses. Increased gonadotropin production can lead to the formation of ovarian cysts [7–9]. Overall, cystic masses are more common than solid masses.

1. Non-Neoplastic Ovarian Cysts
    a. Classification
        i. Cystic masses can be further categorized into simple versus complex [10].
        ii. While simple cysts are comprised of clear fluid, complex cysts may have debris or solid components [10].
        iii. Cystic masses can also be categorized as functional or non-functional [8].
    b. Clinical presentation
        i. Cystic masses may be identified incidentally on ultrasound or present with symptoms including rupture, hemorrhage, or torsion [8].
        ii. Classically, cysts that are symptomatic and >5 cm in size are thought to be at increased risk of torsion.
        iii. Size thresholds for offering elective ovarian cystectomy continue to evolve.
        iv. Neonatal ovarian cysts often regress spontaneously within a year, and observation is recommended [11–13].
2. Ovarian Tumors
    a. Classification
        i. Ovarian tumors are classified by the World Health Organization (WHO) classification system, which distinguishes by the cell of origin [14].
        ii. The types of tumors include: (1) germ cell tumors, (2) cell surface epithelial tumors, (3) sex cord stromal tumors, (4) miscellaneous/other.
            a) Germ cell tumors originate from pluripotent germ cells [15, 16]. In children, germ cell tumors are the most common ovarian tumor whereas, in adults, epithelial ovarian tumors are the most common [15, 17]. Germ cell tumors are mostly benign and can be further categorized into the following types: (1) mature cystic teratoma, (2) immature teratoma, (3) dysgerminoma, (4) mixed germ cell tumor, (5) yolk cell tumor, (6) and choriocarcinoma [15]. These are typically managed surgically per the Children's Oncology Group (COG) staging guidelines. Current literature supports contralateral ovary and uterus preservation (7) [19].
                (1) Mature (Cystic) Teratoma or Dermoid: These are the most common ovarian neoplasm overall and make up 10–20% of

ovarian tumors [15, 18, 20]. Mature teratomas have at least two germ cell layers. Up to 20% of patients have bilateral tumors [21]. On imaging, these tumors may contain fat and/or calcifications. Overall, these tumors have a good prognosis [22], however, an elevated AFP and/or beta HCG level portends an increased risk of malignancy, and these tumors are associated with a risk of gliomatosis peritonei.

(2) Immature (cystic) teratomas are notable for containing embryonal tissue and solid components. Infants and children with immature teratomas typically present with a large palpable mass. Similar to mature teratomas, immature teratomas will also have elevated AFP and/or beta HCG levels and are associated with a risk of gliomatosis peritonei [15].

(3) Dysgerminomas are the most common malignant ovarian tumor [15]. Up to 15% of patients have bilateral tumors [8, 23]. Dysgerminomas are more prevalent in adolescents and patients with gonadal dysgenesis. Features associated with dysgerminomas include elevated LDH and hypercalcemia (associated with parathyroid hormone-related peptide release from a paraneoplastic syndrome) [17]. On imaging, dysgerminomas have the following characteristic features: a solid mass with regions of necrosis (with or without hemorrhage), foci of speckled calcifications, and hypoechoic fibromuscular septa.

(4) Mixed Germ cell tumors: The clinical presentation depends on the components. The components of the germ cell tumor dictate the tumor marker elevation.

(5) Yolk Sac Tumor/Endodermal sinus tumors are the second most common ovarian tumor overall in children [17]. Yolk sac tumors are comprised of solid and cystic components. These tumors have elevated AFP and LDH levels and are aggressive in nature [17].

(6) Embryonal Cell Tumors are solid. Patients with embryonal cell tumors may present with precocious puberty and have elevations of beta HCG [17].

(7) Choriocarcinomas are similar to embryonal cell tumors, these are also solid tumors that may present with precocious puberty. They are associated with elevated beta HCG levels.

b) Cell Surface epithelial tumors: These are the most common type of ovarian tumors in adults [24]. The types of surface epithelial tumors include: (1) serous cystadenoma, (2) mucinous cystadenoma, (3) borderline tumors, and (4) cystadenocarcinoma [15].

(1) Serous cystadenomas are the most common type of surface epithelial tumors and are generally benign [15]. They are unilocular and do not have septations. In up to 20% of patients,

they are bilateral [25]. Patients with serous cystadenomas may have elevated Ca 125 levels.

(2) Mucinous cystadenomas unlike serous cystadenomas, have septations and are generally unilateral [15]. They can grow to be extremely large. About 80% of mucinous tumors are benign with 10% being borderline and another 10% malignant. Patients with mucinous cystadenomas may also have elevated Ca 125 levels.

(3) Borderline ovarian tumors are more common in adolescents than children. They have papillary projections and do not invade the stroma. Patients with borderline ovarian tumors may have elevated Ca 125 levels. Borderline ovarian tumors overall have a good prognosis.

c) Sex cord–stromal tumors: There are multiple types of sex cord tumors including: (1) Juvenile granulosa cell tumor, (2) Sertoli–Leydig cell tumors, (3) Sex cord tumor with tubular annules, (4) Fibroma, (5) Thecoma, (6) Sclerosing stromal tumors, (7) Steroid cell tumors. In general, there is a higher risk of feminizing tumors in children <9 years old and virilizing tumors in children >9 years old [26].

(1) Juvenile Granulosa Tumors are estrogen-producing tumors with high inhibin A levels and low gonadotropin levels [15, 26, 27]. Children with granulosa tumors present with precocious puberty. These tumors are solid with a cystic and/or hemorrhagic component.

(2) Sertoli–Leydig Tumors are testosterone-producing tumors with low gonadotropin levels [15, 27]. Children with Sertoli–Leydig tumors frequently present with amenorrhea or virilization. Sertoli–Leydig tumors are heterogenous histologically with solid and cystic components. They may have elevated AFP and/or Ca 125 levels.

(3) Sex cord tumor with Tubular Annules: These are calcified tumors associated with Peutz–Jeghers syndrome. Medical management is the cornerstone of treatment of sex cord tumors with tubular annules.

(4) Fibromas are rare but often benign tumors [27]. These tumors are generally bilateral [8]. They may be associated with basal cell nevus syndrome (PTCH gene mutation). Children with fibromas may develop amenorrhea.

(5) Thecomas can grow to a large size [8, 27]. Luteinized thecomas are at risk for progression to sclerosing peritonitis [8].

(6) Sclerosing stromal tumors may secrete estrogen. Sclerosing stromal tumors are more frequent in patients with irregular menstrual cycles [27].

        (7) Steroid cell tumors are rare in children. They secrete steroid hormones, and they are associated with increased testosterone, increased urinary 17-ketosteroid, and low gonadotropins. These tumors can cause androgenic pseudo precocity.

    d) Other

        (1) Gonadoblastoma: Children with Turner syndrome are at increased risk of developing gonadoblastoma. These are associated with disorders of sexual development and are thought to represent a carcinoma-in-situ. These elements can progress to various germ cell tumors such as yolk sac or choriocarcinoma with a high risk of malignancy [28].
        (2) Germ cell–Sex Cord Stromal Tumors have both germ cell and sex cord elements. About 10% of these will contain malignant germ cell elements. The clinical presentation includes amenorrhea, delayed menarche, and/or virilization [16].
        (3) Small Cell Carcinoma of the ovary is a rare aggressive tumor that is similar to malignant rhabdoid tumors [15]. Small cell carcinoma of the ovary may be associated with hypercalcemia. Grossly, this tumor is a solid mass with cystic components. These tumors have a poor prognosis.

C. Clinical Features

1. Symptoms: Ovarian masses may not have any associated symptoms [7]. Symptomatic patients may experience a palpable mass, abdominal pain (may also be secondary to associated torsion), abdominal distension, and precocious puberty (more commonly seen in juvenile granulosa cell tumors or some Sertoli–Leydig tumors) [5, 14, 15]. Patients may also experience symptoms of mass effects such as nausea, vomiting, urinary frequency, or urgency [14].
2. History: In adolescent patients, it is important to ask about menstrual cycle history, sexual history, and chronic pelvic pain. In adolescents with chronic pelvic pain, endometriosis should be considered in the differential diagnosis [8].

D. Examination:

1. Abdominal exam: On an abdominal exam, one may be able to palpate a large ovarian mass [8].
2. Breast and axilla exam: One should perform a breast exam and evaluate for axillary hair to assess for precocious puberty [8].
3. Genitourinary exam: In children, one should perform an evaluation for pubic hair for stigmata of precocious puberty [8]. For children, a bimanual exam may be performed via the rectum [8, 29]. For adolescents who are sexually active, a bimanual exam may be performed vaginally [8, 29].

E. Diagnosis

1. Laboratory Tests

   a. Hormone levels: The following hormones may be elevated in patients with functional ovarian masses: (1) Estradiol, (2) Testosterone, (3) Urinary 17-ketosteroid, and (4) gonadotropins [14]

      i. Granulosa cell tumor: Estradiol (high), testosterone (high), gonadotropins (low) [7]
      ii. Sertoli–Leydig tumor: Estradiol (high), testosterone (high) [7]
      iii. Gonadoblastoma: Estradiol (high), testosterone (high), urinary 17-ketosteroid (high), gonadotropins (low) [14]

   b. Tumor markers: Tumor markers should be drawn as they may be elevated in patients with malignant ovarian masses: (1) Lactate dehydrogenase (LDH), (2) beta-human chorionic gonadotropin (beta HCG), (3) Cancer antigen 19-9 (CA 19-9), (4) Inhibin A, (5) CA 125, and (6) carcinoembryonic antigen (CEA) [14, 15, 30, 31]. It is important to note that some malignant tumors will not have an elevation of tumor markers (Table 1).
   c. Thyroid hormone levels (TSH, T4): Hypothyroidism may lead to the development of bilateral ovarian cysts that resolve with thyroid replacement [13].
   d. Pregnancy test: A pregnancy test should be obtained in adolescent patients [13].
   e. Cancer predisposition syndromes: In patients with the following ovarian tumors, one should consider work-up for the associated cancer predisposition syndromes [15].

      a. Juvenile granulosa cell tumor: Ollier disease, Maffucci syndrome
      b. Sertoli–Leydig cell tumors (type of germ cell sex cord tumor): Ollier disease and DICER1 syndrome (higher risk for pleuropulmonary blastoma, Wilms tumor, rhabdomyosarcoma, thyroid cancer, etc.) [15, 32]

**Table 1** Elevated tumor markers seen in ovarian tumors

| AFP | Beta HCG | Inhibin A | LDH | CA 125 | CA 19-9 | CEA |
|---|---|---|---|---|---|---|
| Yolk sac tumor<br>Immature teratoma<br>Embryonal carcinoma<br>Mixed germ cell tumor,<br>Sertoli–Leydig tumors | Choriocarcinoma<br>Embryonal Carcinoma<br>Dysgerminoma | Juvenile Granulosa Cell tumor | Dysgerminoma<br>Germ cell tumor | Epithelial tumor<br>Endometrioma<br>Dysgerminoma<br>Sertoli–Leydig tumor | Epithelial tumor | Epithelial tumor |

    c. Gonadoblastoma: WT-1 related disorders, Turner syndrome
    d. Sex cord tumor with annular tubules: Peutz–Jeghers syndrome
    e. Small cell carcinoma: Rhabdoid tumor predisposition syndrome

2. Radiographic Studies:

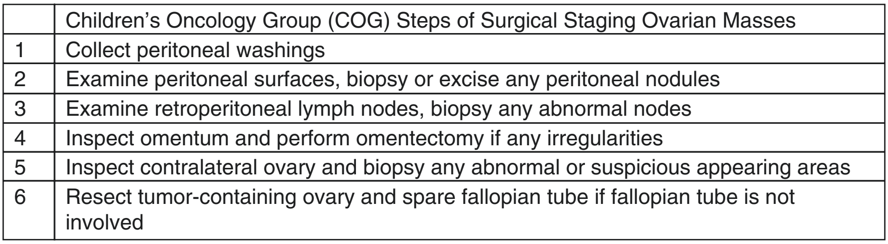
    a. Ultrasound is considered the first-line imaging for ovarian masses [33]. The features to evaluate on ultrasound include free fluid, mass size, solid/cystic, presence of septations, uterus size, endometrial thickness, ovary size, presence of follicles in the ovary, and the presence of blood flow to the ovary (vascular flow pattern and arterial waveform with doppler) [30]. Specific features concerning malignancy include size >8–10 cm, large tumor volume >184 cc (measured with the prolate ellipsoid formula), and solid components [7, 15, 34]. There are multiple tools that have been developed to assess ovarian masses using ultrasound findings, including the DePriest score and the Ueland index [7, 15, 35–37].

        i. DePriest score is a tool used to distinguish benign and malignant ovarian masses [7, 35]. This was primarily studied in the adult population. The score is comprised of (1) tumor volume, (2) cyst wall structure, and (3) septa structure with scores for each component from 0 to 4 (total score of 12). A DePriest score <5 was associated with benign mass. The Ueland index modified the DePriest scoring system and was similarly studied in the adult population [7, 36, 37].
        ii. The Ueland index consists of (1) tumor volume and (2) tumor structure with scores for each component from 0 to 5 (total score 10). A score >7 is concerning for malignancy [7, 36, 37].
        iii. The ovarian crescent sign is visualized when there is a rim of healthy tissue around the ovary that has a benign mass [7].

    b. MRI has been evaluated in small studies, MRI can be effective to distinguish between benign and malignant masses [1, 33]. The advantages of MRI are that it provides better soft-tissue visualization, specifically for differentiating ovarian and uterine masses, and limits radiation exposure [14, 33, 38]. Features on MRI associated with malignancy include size >4 cm, profuse papillary projections, areas of necrosis, "thicker walls [>3 mm], presence of septa and/or vegetation within the mass, increased signal intensity on T2-weighted imaging, increased contrast enhancement, ascites, peritoneal implants, and bilaterality" [1].
    c. CT of the abdomen and pelvis may be helpful if the ultrasound is inconclusive.
    d. CT chest is important to obtain for staging a malignancy.

F. Treatment

1. Initial Measures: Prior to initiation of chemotherapy for large germ cell tumors, a biopsy should be obtained [24].
2. Subsequent Measures: Chemotherapy can be considered for Stages 2–4 germ cell tumors (cisplatin, etoposide, and bleomycin) and for advanced-stage sex cord–stromal (bleomycin, etoposide, and cisplatin) and epithelial tumors (cisplatin, cyclophosphamide, and paclitaxel) [17, 39].
3. Preoperative Preparation:
    a. Ensure that appropriate bloodwork has been drawn pre-operatively if there is a concern for malignancy based on radiographic features and clinical suspicion. Tumors markers after resection may not be reliable.
    b. If not an acute presentation, cross-sectional imaging in a stable patient can aid in surgical planning.
    c. An open approach should be considered for larger tumors (>10 cm) to avoid tumor spillage.
    d. Foley catheter placement for bladder decompression is typical.
4. Operative Treatment: Operative management should be considered for (1) enlarging or symptomatic ovarian cyst, especially if >5 cm, (2) complex mass with concerning features ("elevated tumor markers, precocious puberty, size >8 cm, features [solid component, papillary projections, ill-defined borders, ascites, thick septations, extension into surrounding structures, metastatic disease], or (3) history of ovarian malignancy" [30]. Some studies advocate the removal of ovarian cysts >8 cm [11].
    a. Laparoscopic versus open: There is an increasing adoption and preference for a laparoscopic approach for ovarian tumors; however, one may need to convert to open (lower midline versus Pfannenstiel incision) to prevent tumor spillage or resect the entirety of a large ovarian mass [24].
    b. Role of frozen section: Frozen section is considered inadequate for pediatric ovarian tumors, with lacking data in the pediatric population [7]. Presently, there is an increased discrepancy in agreement between frozen and permanent for borderline, purely cystic tumors, and mucinous tumors [7, 40–42].
    c. Surgical Staging: Surgical staging of ovarian tumors has been described by the Children's Oncology Group [17, 43–45]. The steps are outlined below [17, 43–45] (Fig. 1; Table 2):
    d. FIGO Surgical Staging for Epithelial tumors: Epithelial ovarian tumors use the FIGO staging while germ cell tumors use the Children's Oncology Group staging [46]. The FIGO Staging is demonstrated below (Table 3):
    e. Cystectomy: Recommended for ovarian cysts, and preferable to oophorectomy, even large ovarian cysts [7, 13]; There are risks to oophorectomy, specifically a higher risk of premature ovarian failure and early menopause even with a unilateral oophorectomy [2, 22, 47]. Excision is associated with a lower rate of recurrence compared to aspiration or fenestration



| | Children's Oncology Group (COG) Steps of Surgical Staging Ovarian Masses |
|---|---|
| 1 | Collect peritoneal washings |
| 2 | Examine peritoneal surfaces, biopsy or excise any peritoneal nodules |
| 3 | Examine retroperitoneal lymph nodes, biopsy any abnormal nodes |
| 4 | Inspect omentum and perform omentectomy if any irregularities |
| 5 | Inspect contralateral ovary and biopsy any abnormal or suspicious appearing areas |
| 6 | Resect tumor-containing ovary and spare fallopian tube if fallopian tube is not involved |

**Fig. 1** Children's Oncology Group (COG) steps of surgical staging ovarian masses

**Table 2** COG staging system

| COG staging system | |
|---|---|
| Stage 1 | Tumor entirely removed, normal tumor markers |
| Stage 2 | Microscopic traces of tumor, tumor markers do not return to normal after surgery |
| Stage 3 | Gross tumor left behind, positive lymph nodes |
| Stage 4 | Metastatic spread of tumor |

**Table 3** FIGO surgical staging system

| FIGO surgical staging system | |
|---|---|
| Stage 1 | Tumor confined to ovaries or fallopian tubes |
| Stage 2 | Tumor involves 1 or both ovaries or fallopian tubes with pelvic extension or primary peritoneal cancer |
| Stage 3 | Tumor involves 1 or both ovaries or fallopian tubes, or primary peritoneal cancer, with cytologically or histologically confirmed spread to the peritoneum outside of the pelvis and/or metastasis to the retroperitoneal lymph nodes |
| Stage 4 | Distant metastasis (excludes peritoneal metastases) |

only [8]. Ovarian cysts can be excised by incising along antimesenteric border of the ovary to visualize the entirety of the cyst wall. This can then be shelled out in its entirety, or the posterior cyst wall may be left in place if technically unable to be separated. The risk of recurrence is significantly higher if fenestration without full excision is performed [13].

f. Partial Oophorectomy: For mature germ cell tumors or masses that are localized with negative markers or benign appearing radiographic features, ovarian conserving resection should be performed. Typically, the ovarian parenchymal edges do not require re-approximation after ovarian-sparing tumor resection [7, 17]. If pathology confirms a malignancy, then return to the operating room for completion of oophorectomy may be indicated.

g. Oophorectomy: Oophorectomy with or without salpingectomy is indicated for ovarian neoplasms with malignant features. For germ cell tumors with malignant features, there is evidence to support contralateral ovary and uterus preservation [7, 45]. In general, if the tumor does not invade the hilum or invade the fallopian tube, one should perform an oophorectomy alone [17]. However, if the fallopian tube is involved in germ cell tumors, removal of the fallopian tube is recommended [7, 17].

h. Contralateral ovarian biopsy: This is not recommended for germ cell tumors [7]. Contralateral ovarian biopsy may be considered if there are visible abnormalities of the contralateral ovary [7].
i. Lymph node biopsy: For germ cell tumors, one should only perform lymph node biopsy for abnormal lymph nodes or enlarged lymph nodes (>2 cm) [17]. For epithelial tumors, ipsilateral lymph node dissection should always be performed [17].
j. Ovarian preserving surgery/Fertility sparing approach: Ovarian preserving surgery involves the resection of abnormal tissue and leaving healthy ovarian tissue behind. This may be utilized in "ovarian cystectomy, detorsion, and/or drainage, fenestration, or unroofing of ovarian cysts" [2, 30]. This is also the preferred approach for (1) enlarging or symptomatic cysts (2) complex masses with negative tumor markers and no concerning features (3) germ cell tumors, (4) low grade epithelial ovarian tumors, and (5) borderline ovarian tumors [7, 30] (Fig. 2). Additionally, ovarian cryopreservation may be considered for pre-pubertal children who require unilateral or bilateral oophorectomies [22, 48]. This approach involves laparoscopic unilateral oophorectomy followed by cryopreservation and subsequent reimplantation after treatment has been completed [48].

5. Postoperative Treatment: Complications associated with surgery for ovarian masses include tumor spill, peritonitis, bleeding, infection, postoperative hernia, and damage to adjacent structures (bladder, rectum, and ureter) [14].
6. Postoperative Surveillance: Following surgery, infants and children with ovarian neoplasms need to be monitored closely. Specifically, they should initially be monitored with imaging (ultrasound, CT abdomen/pelvis, or MRI) and tumor marker surveillance every 3 months for the first 3 years and then yearly [13].

## Additional Notes

The patient presenting with acute torsion:

Although unusual in the case of an ovarian malignancy, patients may present acutely with torsion. This can complicate the diagnostic work-up as the tumor markers will not be available and there may not be time for additional cross-sectional imaging. Malignant ovarian masses are far less likely to present with torsion compared to benign ovarian masses; however, when this occurs, a staged approach is indicated [5]. If a discrete mass can be shelled out of the ovarian parenchyma in an ovarian-preserving manner, this should be performed with care taken to avoid rupture and spillage. If the ovary and mass are significantly congested and edematous

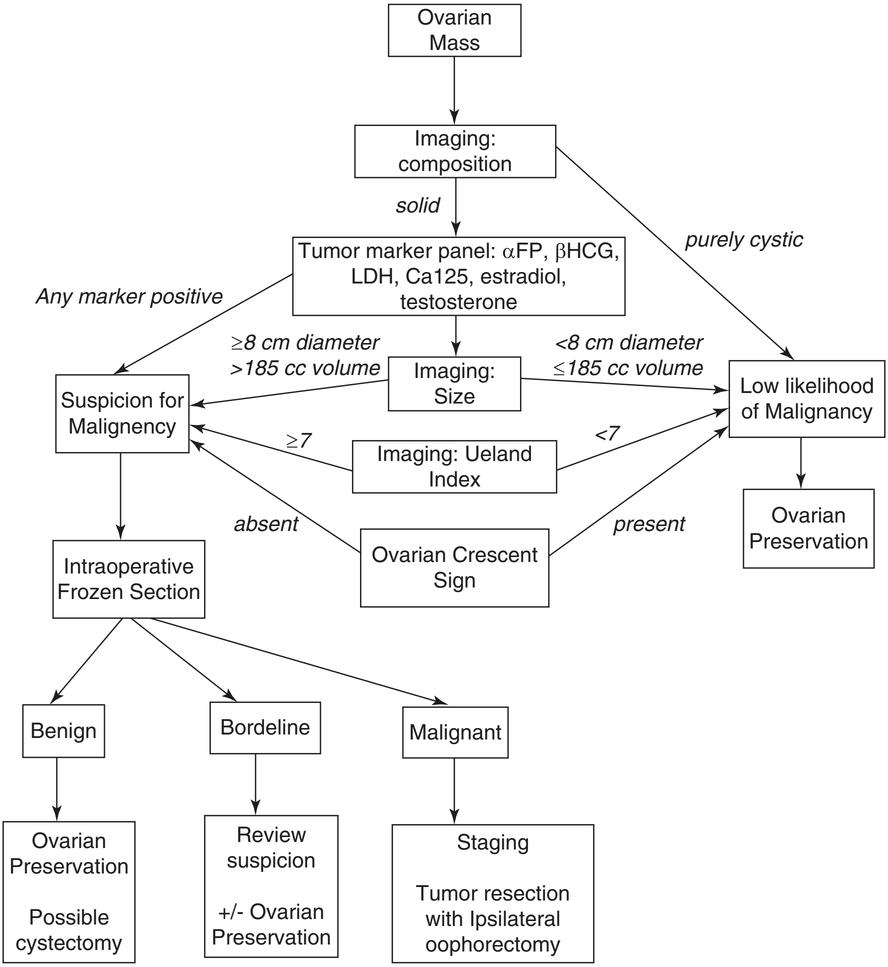

Ovarian Mass
Imaging: composition
solid
purely cystic
Tumor marker panel: αFP, βHCG, LDH, Ca125, estradiol, testosterone
Any marker positive
≥8 cm diameter >185 cc volume
Imaging: Size
<8 cm diameter ≤185 cc volume
Suspicion for Malignency
Low likelihood of Malignancy
≥7
Imaging: Ueland Index
<7
absent
Ovarian Crescent Sign
present
Ovarian Preservation
Intraoperative Frozen Section
Benign
Bordeline
Malignant
Ovarian Preservation
Possible cystectomy
Review suspicion
+/- Ovarian Preservation
Staging
Tumor resection with Ipsilateral oophorectomy

**Fig. 2** Treatment algorithm for pediatric ovarian mass developed by Renaud et al (Reprinted with permission from Elsevier and Dr. Elizabeth Renaud et al. [7])

such that mass excision is not feasible, ovarian detorsion should be performed and the surgery concluded. Tumor markers should be sent prior to excision and subsequent follow-up imaging obtained within 4–6 weeks. Surgical resection can then be pursued in a non-emergent fashion to optimize the chance of ovarian preservation for masses that ultimately are benign and completion oophorectomy can be performed where indicated for malignant masses.

Laparoscopic or open approach for large masses:

Minimally invasive techniques are increasingly considered the gold standard when approaching ovarian lesions. Simple cystic lesions with no concerning radiographic features can be evaluated laparoscopically and cystectomy performed with removal facilitated by controlled decompression of the fluid. This is performed either with laparoscopic needle decompression or by placing the cyst in a laparoscopic bag to open and decompress with a suction device. Complex lesions or masses that contain solid components that are consistent with mature teratomas can be similarly approached and removed intact within a laparoscopic specimen retrieval bag. For malignant lesions, the decision to proceed laparoscopically following abdominal evaluation and staging protocols should be tailored to the surgeon's comfort with minimally invasive techniques. The goal of complete resection without spillage should not be compromised by pursuing a laparoscopic approach. For larger tumors (>10 cm), consideration of a Pfannenstiel incision or lower midline incision may be indicated. A Pfannenstiel incision can be made to just fit the tumor size thus not endangering an intact tumor removal but permitting a faster, less painful recovery, and improved cosmesis.

Management of an ovarian germ cell tumor invading surrounding pelvic structures:

If intra-operative evaluation reveals that the ovarian mass is closely adherent to adjacent pelvic organs, such as uterus, rectum, colon, or bladder, there is little benefit to attempting a complete resection [45]. A complete pelvic exenteration is not recommended as adjuvant chemotherapy has demonstrated improved outcomes while preserving function, limiting associated morbidities, and preserving future fertility. A careful laparoscopic or open staging should occur intra-operatively including obtaining peritoneal washings with assessment and biopsy of any abnormalities of peritoneum, omentum, and contralateral ovary. The mass itself should be biopsied and the operation concluded with plans to return following adjuvant chemotherapy for surgical excision and local control.

## Study Questions

Question 1: What is a component of the Children's Oncology Group (COG) intra-operative staging protocol for germ cell tumors?

a. Unilateral lymph node sampling
b. Examination of inguinal lymph nodes
c. Bilateral salpingectomy
d. Omental inspection and omentectomy if abnormal

Answer to Question 1: (d.): Omental inspection and omentectomy if abnormal. The COG intra-operative staging protocol involves the following: (1) Collect peritoneal washings, (2) Examine peritoneal surfaces, biopsy or excise any peritoneal nodules, (3) Examine retroperitoneal lymph nodes, biopsy abnormal or enlarged (>2 cm) lymph nodes (4) Inspect omentum and perform omentectomy, (5) Contralateral ovarian inspection with biopsy of any abnormal lesions, and (6) Resect tumor-containing ovary and spare fallopian tube if fallopian tube is not involved. A is incorrect as lymph node sampling is recommended only for abnormal or enlarged nodes. B is incorrect as retroperitoneal lymph node examination is recommended. C is incorrect as the fallopian should be spared if the fallopian tube is not involved. D is correct as complete omentectomy is recommended if any omental lesions are identified.

Question 2: Which is true of cystic teratomas?

a. Up to half of patients will have bilateral tumors.
b. These tumors have high malignant potential.
c. Imaging may show the presence of fat and/or calcifications.
d. A low AFP level is associated with risk of gliomatosis peritonei.

Answer to Question 2: (c.): Imaging may show the presence of fat and/or calcifications. On imaging, these tumors may contain fat and/or calcifications. Up to 20% of patients have bilateral tumors so A is incorrect. Overall, these tumors have a good prognosis with low malignant potential, therefore B is incorrect. An elevated AFP is associated with immature teratoma and an increased risk of malignancy. The presence of immature elements is also seen more often in the rare cases of gliomatosis peritonei. Therefore, D is incorrect.

Question 3: Match the appropriate type of ovarian tumor with the expected tumor marker elevation.

| Epithelial tumor | B HCG |
|---|---|
| Yolk Sac tumor | CA125 |
| Choriocarcinoma | AFP |
| Dysgerminoma | LDH |

Answer to Question 3: Epithelial cancers are associated with elevated CA 125, CA 19-9, and CEA. Yolk sac tumors have elevated AFP. Choriocarcinomas have elevated B HCG. Dysgerminomas have elevated LDH and CA 125.

## References

1. van Nimwegen LW, Mavinkurve-Groothuis AM, de Krijger RR, Hulsker CC, et al. MR imaging in discriminating between benign and malignant paediatric ovarian masses: a systematic review. Eur Radiol. 2020;30(2):1166–81.
2. Gonzalez DO, Cooper JN, Aldrink JH, Hewitt GD, et al. Variability in surgical management of benign ovarian neoplasms in children. J Pediatr Surg. 2017;52(6):944–50.
3. Skinner MA, Schlatter MG, Heifetz SA, Grosfeld JL. Ovarian neoplasms in children. Arch Surg. 1993;128(8):849–54.
4. Oelschlager AEA, Gow KW, Morse CB, Lara-Torre E. Management of large ovarian neoplasms in pediatric and adolescent females. J Pediatr Adolesc Gynecol. 2016;29(2):88–94.
5. Cass DL, Hawkins E, Brandt ML, Chintagumpala M, et al. Surgery for ovarian masses in infants, children, and adolescents: 102 consecutive patients treated in a 15-year period. J Pediatr Surg. 2001;36(5):693–9.
6. Stein E. Anorectal and colon diseases: textbook and color atlas of proctology; with 54 tables. Berlin: Springer Science and Business Media; 2003.

7. Renaud EJ, Sømme S, Islam S, Cameron DB, et al. Ovarian masses in the child and adolescent: an American Pediatric Surgical Association outcomes and evidence-based practice committee systematic review. J Pediatr Surg. 2019;54(3):369–77.
8. Kelleher CM, Goldstein AM. Adnexal masses in children and adolescents. Clin Obstet Gynecol. 2015;58(1):76–92.
9. Brandt ML, Helmrath MA. Ovarian cysts in infants and children. Seminars Pediatr Surg. 2005;14:78–85.
10. Laufer MR. Ovarian cysts in infants, children, and adolescents. 2020. www.uptodate.com/contents/ovarian-cysts-in-infants-children-and-adolescents#!
11. Hayes-Jordan A. Surgical management of the incidentally identified ovarian mass. Seminars Pediatr Surg. 2005;14(2):106–10.
12. Dolgin SE. Ovarian masses in the newborn. Seminars Pediatr Surg. 2000;9(Part 3):121–7.
13. Holcomb GW III, Murphy HP, St. Peter SD. Holcomb and Ashcraft's pediatric surgery. 7th ed. New York: Elsevier; 2020. p. 211–89.
14. Ovarian Tumors. American Pediatric Surgical Association and Unbound Medicine; 2020 [updated November 25th, 2020; cited May 10th, 2021]. Available from: https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829126/all/Ovarian_Tumors
15. Lala SV, Strubel N. Ovarian neoplasms of childhood. Pediatr Radiol. 2019;49(11):1463–75.
16. Carcangiu M, Kurman RJ, Carcangiu ML, Herrington CS. WHO classification of tumours of female reproductive organs. Lyon: International Agency for Research on Cancer; 2014.
17. Germ Cell Ovarian Tumors. American Pediatric Surgical Association and Unbound Medicine; 2020 [updated June 17, 2018; cited May 10th, 2021]. Available from: https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829126/all/Ovarian_Tumors
18. Kim MJ, Kim NY, Lee D, Yoon B, Choi D. Clinical characteristics of ovarian teratoma: age-focused retrospective analysis of 580 cases. Obstet Gynecol. 2011;205(1):32.e1–4.
19. Newly Diagnosed with Germ Cell Tumors. The Children's Oncology Group; 2011 [cited 6/13/2021]. Available from: https://www.newly-diagnosed-with-germ-cell-tumors#:~:text=COG%20Germ%20Cell%20Tumor%20Staging,return%20to%20normal%20following%20surgery
20. Bloomfield T. Benign cystic teratomas of the ovary: a review of seventy-two cases. Eur J Obstet Gynecol Reproduct Biol. 1987;25(3):231–7.
21. Taskinen S, Urtane A, Fagerholm R, Lohi J, Taskinen M. Metachronous benign ovarian tumors are not uncommon in children. J Pediatr Surg. 2014;49(4):543–5.
22. Braungart S, Craigie RJ, Farrelly P, Losty PD, Surgeons CCLG, Collaborators. Operative management of pediatric ovarian tumors and the challenge of fertility-preservation: results from the UK CCLG Surgeons Cancer Group Nationwide Study. J Pediatr Surg. 2020;55(11):2425–9.
23. Von Allmen D. Malignant lesions of the ovary in childhood. Seminars Pediatr Surg. 2005;14(2):100–5.
24. Oophorectomy for Tumor. American Pediatric Surgical Association and Unbound Medicine; 2020 [updated January 31, 2017; cited May 10th, 2021]. Available from: https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829237/all/Oophorectomy_for_Tumor
25. Lack E, Young R, Scully R. Pathology of ovarian neoplasms in childhood and adolescence. Pathol Annu. 1992;27:281–356.
26. Stocker JT, Dehner LP. Pediatric pathology. Philadelphia, PA: Lippincott Williams & Wilkins; 2001.
27. Sex Cord-Stromal Ovarian Tumors. American Pediatric Surgical Association and Unbound Medicine; 2020 [updated August 21, 2017; cited May 10th, 2021]. Available from: https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829126/all/Ovarian_Tumors
28. Schultz KAP, Sencer SF, Messinger Y, Neglia JP, Steiner ME. Pediatric ovarian tumors: a review of 67 cases. Pediatr Blood Cancer. 2005;44(2):167–73.
29. Templeman CL, Fallat ME. Benign ovarian masses. Seminars Pediatr Surg. 2005;14(2):93–9.
30. Aldrink JH, Gonzalez DO, Sales SP, Deans KJ, et al. Using quality improvement methodology to improve ovarian salvage for benign ovarian masses. J Pediatr Surg. 2018;53(1):67–72.

31. Lawrence AE, Fallat ME, Hewitt G, Hertweck P, et al. Understanding the value of tumor markers in pediatric ovarian neoplasms. J Pediatr Surg. 2020;55(1):122–5.
32. Schultz KAP, Williams GM, Kamihara J, Stewart DR, et al. DICER1 and associated conditions: identification of at-risk individuals and recommended surveillance strategies. Clin Cancer Res. 2018;24(10):2251–61.
33. Emil S, Youssef F, Arbash G, Baird R, et al. The utility of magnetic resonance imaging in the diagnosis and management of pediatric benign ovarian lesions. J Pediatr Surg. 2018;53(10):2013–8.
34. Abbas PI, Elder SC, Mehollin-Ray AR, Braverman RM, et al. Ovarian lesion volumes as a screening tool for malignancy in adolescent ovarian tumors. J Pediatr Surg. 2015;50(11):1933–6.
35. DePriest P, Shenson D, Fried A, Hunter J, et al. A morphology index based on sonographic findings in ovarian cancer. Gynecol Oncol. 1993;51(1):7–11.
36. Ueland F, DePriest P, Pavlik E, Kryscio R, van Nagell JJ. Preoperative differentiation of malignant from benign ovarian tumors: the efficacy of morphology indexing and Doppler flow sonography. Gynecol Oncol. 2003;91(1):46–50.
37. Stankovic Z, Bjelica A, Djukic M, Savic D. Value of ultrasonographic detection of normal ovarian tissue in the differential diagnosis of adnexal masses in pediatric patients. Ultrasound Obstet Gynecol. 2010;36(1):88–92.
38. Baker PM, Oliva E. Immunohistochemistry as a tool in the differential diagnosis of ovarian tumors: an update. Int J Gynecol Pathol. 2005;24(1):39–55.
39. Raju S, Neglen P. Popliteal vein entrapment: a benign venographic feature or a pathologic entity? J Vasc Surg. 2000;31(4):631–41.
40. Song T, Choi CH, Lee Y, Kim T, et al. Pediatric borderline ovarian tumors: a retrospective analysis. J Pediatr Surg. 2010;45(10):1955–60.
41. Gol M, Baloglu A, Yigit S, Dogan M, et al. Accuracy of frozen section diagnosis in ovarian tumors: is there a change in the course of time? Int J Gynecol Cancer. 2003;13(5):593.
42. Pinto PB, Andrade LA, Derchain SF. Accuracy of intraoperative frozen section diagnosis of ovarian tumors. Gynecol Oncol. 2001;81(2):230–2.
43. Ramli R, Mansor H, Abdullah Y, Mulalek N, Yusoff WAB. Laparoscopic detorsion of gangrenous adnexae followed by interval cystectomy: case report and review of the literature. Int J Reprod Contracept Obstet Gynecol. 2015;4(3):899.
44. Styer AK, Laufer MR. Ovarian bivalving after detorsion. Fertil Steril. 2002;77(5):1053–5.
45. Billmire D, Vinocur C, Rescorla F, Cushing B, et al. Outcome and staging evaluation in malignant germ cell tumors of the ovary in children and adolescents: an intergroup study. J Pediatr Surg. 2004;39(3):424–9.
46. Prat J. Ovarian, fallopian tube and peritoneal cancer staging: rationale and explanation of new FIGO staging 2013. Best Pract Res Clin Obstet Gynaecol. 2015;29(6):858–69.
47. Yasui T, Hayashi K, Mizunuma H, Kubota T, et al. Factors associated with premature ovarian failure, early menopause and earlier onset of menopause in Japanese women. Maturitas. 2012;72(3):249–55.
48. Rowell EE, Corkum KS, Lautz TB, Laronda MM, et al. Laparoscopic unilateral oophorectomy for ovarian tissue cryopreservation in children. J Pediatr Surg. 2019;54(3):543–9.

# Ovarian Torsion

**Muriel A. Cleary**

**Key Points**

1. The presence of Doppler flow on ultrasound imaging does not rule out the presence of ovarian torsion, thus making it a truly clinical diagnosis for which one should have a high index of suspicion.
2. Ovarian sparing surgery with laparoscopic detorsion alone or detorsion with cystectomy is considered the gold standard.
3. The risk of an associated malignancy presenting with ovarian torsion is a low, particularly when the mass at presentation is <8 cm.
4. Follow-up ultrasound surveillance after detorsion or detorsion with cystectomy should be performed 2–3 months following surgery.

## Epidemiology

A. Incidence

   a. National in-patient data from a 2006 KID analysis demonstrates that idiopathic ovarian torsion in the pediatric population is a rare occurrence with an estimated incidence of 4.9:100,000 [1].
   b. A mean age of 14.5 years is reported with peak incidence during early adolescence and immediate post-menarchal years [2].
   c. Children less than 18 years of age constitute about 15% of all ovarian torsion cases reported in larger series [1, 2].

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B. Risk Factors

a. The majority of torsion events occur in women of childbearing age between the ages of 20 and 40 years [3].

b. The presence of an ovarian cyst/mass greater than 5 cm in size has been reported to be a risk factor predisposing to torsion. Reportedly, the pendulous weight of the associated ovary can cause it to flip upon itself around the vascular pedicle. Several case series have shown that the majority of torsion cases involve lesions of 5 cm or greater particularly in the post-menarchal adolescent age group [4–6].

## Relevant Anatomy

A. Anatomy

a. The adnexal structures are encased in peritoneal folds and supported by the suspensory elements of the (1) broad ligament which include the mesometrium, the mesovarium, and the mesosalpinx; (2) the suspensory ligament which contains the ovarian artery; and (3) the utero-ovarian ligament which connects the ovary and the uterus [7] (Fig. 1).

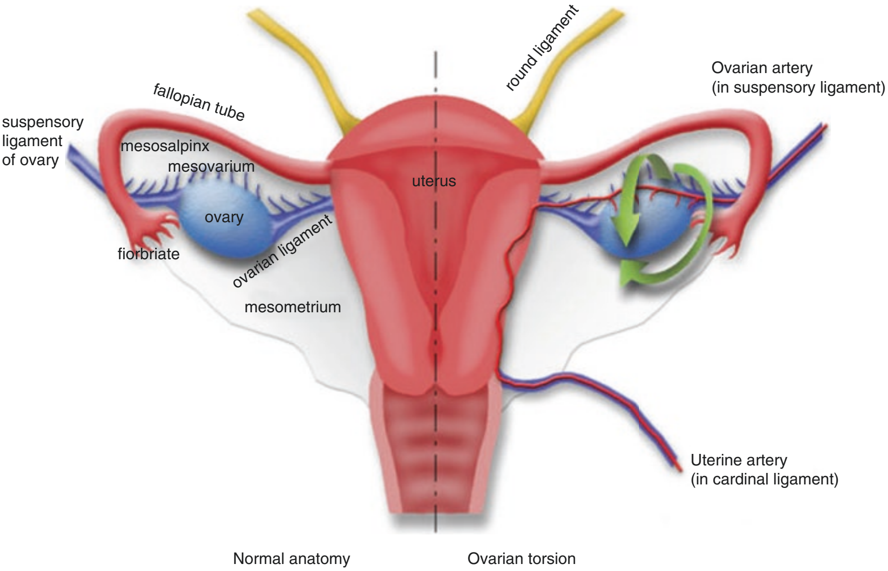
round ligament
Ovarian artery
(in suspensory ligament)
fallopian tube
suspensory
ligament
of ovary
mesosalpinx
mesovarium
uterus
ovary
fiorbriate
ovarian ligament
mesometrium
Uterine artery
(in cardinal ligament)
Normal anatomy
Ovarian torsion

**Fig. 1** Schematic diagram featuring the normal anatomy of adnexal ligaments (left side) and the mechanisms involved in ovarian torsion (green arrow) and in the adnexal torsion (large arrow). Source: Nanni M., Merola M.G., Ianniello S., Orazi C., Schingo P.M., Trinci M. (2016) Ovarian Torsion. In: Miele V., Trinci M. (eds) Imaging Non-traumatic Abdominal Emergencies in Pediatric Patients. Springer, Cham. With cited thanks to Edoard Saperi

- b. The blood supply to the ovary is directed from the ovarian artery within the suspensory ligament and a small ovarian branch that comes off the uterine artery. Thus, as a result of the dual arterial supply, arterial flow can still be detected on Doppler imaging if the torsion is less than 360° and does not cause compression of both the lateral suspensory ligament and the medial utero-ovarian ligament [8].
- c. Adnexal twisting may occur along the mesovarium or the mesosalpinx and be isolated ovarian or fallopian tube torsion events, however, the majority of torsions involve the entire adnexa [7].
- d. The right side is more frequently affected and this has been attributed to the fixed space-occupying nature of the sigmoid colon on the left being somewhat protective compared with the mobile cecum on the right [9].

B. Physiology

- a. Torsion causes impaired outflow of low pressure venous and lymphatic systems and results in edema and adnexal enlargement. Ongoing venous obstruction and engorgement result in worsening compression of the vascular hilum.
- b. In cases of complete torsion (>360°), this will then progress to obstructed arterial flow and resultant ischemia, hemorrhagic infarction, and necrosis [7]. In this scenario, ovaries will take on a blue-black appearance upon exploration.

## Clinical Features

A. Timing

- a. Ovarian torsion is a clinical diagnosis that is supported by history, physical exam findings, and radiographic information. Normal Doppler flow imaging cannot rule out the presence of torsion in a patient with ongoing symptoms [10].
- b. The presenting symptoms can be quite variable and challenging to interpret which commonly results in delay in diagnosis.
- c. The exact duration of time that results in ovarian parenchymal ischemia and necrosis is unknown as there is good collateral blood supply to the ovary. However, delays in treatment and intervention can result in loss of ovarian tissue and potentially impaired fertility [4].
- d. A high degree of suspicion is needed to diagnose ovarian torsion and, once suspected, it requires emergent intervention. The 2019 American College of Obstetricians and Gynecologists Committee Opinion on adnexal torsion in adolescents states that given the importance of ovarian preservation in this young cohort, a negative finding at laparoscopy is an acceptable clinical outcome [11].

## Diagnosis

A. Clinical Presentation

   a. Ovarian torsion accounts for up to 2.7% of all cases of acute abdominal pain in children [2].
   b. The vast majority of patients will present with acute onset non-radiating intermittent abdominal pain. One institutional study found that 100% of patients with ovarian torsion presented with abdominal pain with three-quarters of them reporting a subjective pain severity greater than 8 [4].
   c. Associated nausea and emesis are strongly associated with an active torsion with studies reporting these as presenting symptoms in 87–97% of patients with confirmed ovarian torsion [4, 11, 12]
   d. Pain may be transient and recurrent in the case of intermittent ovarian torsion and can present with days or even weeks of waxing–waning episodes [7].
   e. Physical exam findings typically include abdominal tenderness to palpation that may or may not be accompanied by peritonitis. It is unusual for a pelvic mass to be palpated in this age group.
   f. Laboratory testing does not add significantly to the work-up of ovarian torsion however a negative β-hCG is important to document.
   g. An associated leukocytosis is not a specific finding [12, 13].

B. Differential

   a. The differential diagnosis for abdominal pain in a female pediatric patient is broad and can include acute appendicitis, mesenteric adenitis, urinary tract infection or pyelonephritis, constipation, viral gastroenteritis, tubo-ovarian abscess, ruptured hemorrhagic cyst, ectopic pregnancy or intermittent or acute ovarian torsion.
   b. Hemorrhagic cysts occur in post-pubertal patients during the luteal phase of the menstrual cyst and can average 3–3.5 cm in average diameter thus contributing to a relative size discrepancy on ultrasound. In cases of torsion, however, the ovary typically measures >5 cm and has a volume ratio of ≥15 compared to the normal ovary [14].

C. Radiographic Work-up

   a. Ultrasound

      i. Ultrasound (US) is the gold standard imaging modality to evaluate for ovarian torsion. It is a non-invasive and rapid test that can be done in an urgent setting with a high sensitivity and specificity in skilled hands [15].
      ii. The main features to evaluate on US include (1) ovarian size (dimension > 5 cm or volume > 20 cm$^3$) (2) unilateral ovarian enlargement relative to the other side (volume ratio >12) (3) peripherally positioned follicles (4) medialization of the twisted ovary (5) presence or absence of an associated mass with characterization of solid or cystic components (6) absence of venous or arterial blood flow on Doppler [4, 7, 16] (Fig. 2).

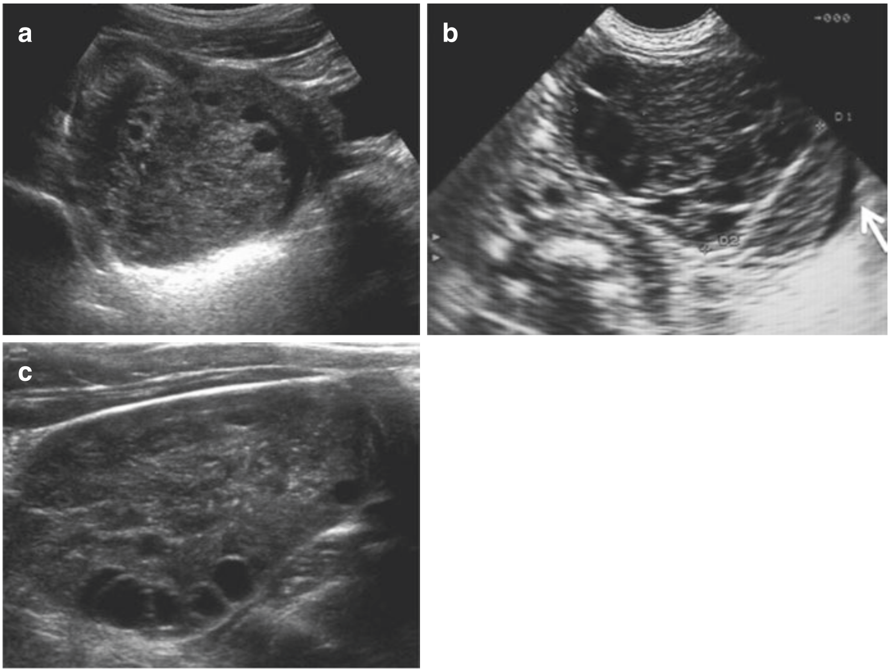

**Fig. 2** Ovarian torsion in an 8-year-old girl. (**a**, **b**) Transverse US shows a unilateral ovarian enlargement, with a central afollicular stroma and a peripheral placement of multiple cortical follicles. (**c**) The enlarged ovary is surrounded by hyperechoic fat and by a thin fluid flap (white arrow). Source: Nanni M., Merola M.G., Ianniello S., Orazi C., Schingo P.M., Trinci M. (2016) Ovarian Torsion. In: Miele V., Trinci M. (eds) Imaging Non-traumatic Abdominal Emergencies in Pediatric Patients. Springer, Cham

iii. Altered venous flow is more common than arterial involvement with as many as two-thirds of patients with proven torsion having documented arterial flow on Doppler [7].
iv. The "whirlpool sign" is a swirling configuration sometimes seen on Doppler that represents the twisted vascular pedicle and carries a sensitivity of 88% and specificity of 87% [17].
v. It is important to note that in a symptomatic patient, the presence of arterial waveforms on Doppler does not rule out the possibility of ovarian torsion with one study citing normal Doppler arterial flow in 60% of surgically confirmed cases of adnexal torsion [18, 19].

b. CT

i. Generally, CT is not recommended as first-line in the work-up for ovarian torsion given the preferred speed and utility of US in this diagnosis without the associated radiation.
ii. A CT can also demonstrate asymmetric ovarian enlargement, central afollicular stroma with peripheralization of follicles, an associated ovarian mass, or the presence of a whirlpool sign. Compared to US, CT is

better at characterizing solid masses and detecting the presence of calcifications in the setting of a teratoma [7, 20]. It may be used for surgical planning when a complex mass is detected.

    iii. A contrast CT can detect abnormal or absent ovarian enhancement however, similar to US, the presence of contrast enhancement does not definitively rule out torsion [12].
    iv. Cross-sectional imaging may shed more light on other gynecological diagnoses such as pyosalpinx, tubo-ovarian abscess, and hemorrhagic and endometriotic ovarian cysts [7, 16]

  c. MRI

    i. MRI is rarely utilized given the high acuity of patients presenting with concern for torsion and its limited availability.
    ii. Use has been described in limited pediatric cases and in older pregnant patients where the ovaries may not be low in the pelvis and are easily visualized on US. Standard protocols include T2, T1-weighed fat-suppressed, and T1 fat-suppressed post-contrast sequences which allow identification of hemorrhage and/or infarction with necrosis and soft-tissue characterization of complex masses [13, 16].
    iii. Radiographic MRI findings are otherwise very similar to those described above in review of CT images [20].

D. Presence of an Ovarian Mass

  a. In premenarchal patients, there is no associated pathology in about 45–50% of cases. In the remaining patients, the majority are simple paratubal or paraovarian cysts or mature teratomas. The most common benign cysts that are reported are physiologic follicular or luteal cysts or serous cystadenomas [4].
  b. Malignancy rates overall are low in the pediatric population and evidence suggests that torsion is not an independent risk factor for malignancy. Therefore, the presence of torsion should not increase one’s suspicion for malignancy [4, 6].
  c. If imaging is suggestive of a complex mixed cystic-solid mass, particularly if it is >8 cm, the literature supports obtaining pre-operative tumor markers including β-hCG, αFP, LDH, CA125, Inhibin A/B [2, 3]. These results will typically not be available at the time of the operative exploration, however, it will aid in confirming a diagnosis post-operatively.

E. Prenatal Diagnosis

  a. Ultrasound remains the modality of choice for the detection and surveillance of neonatal ovarian cysts. Antenatal detection of ovarian cysts is common in the third trimester given excessive maternal and placental hormonal stimulation and is more frequently seen in pregnancies complicated by diabetes mellites, preeclampsia, and Rh isoimmunization [21]. Other differential diagnoses for an intra-abdominal prenatal or neonatal cyst include mesenteric cyst, lymphatic malformation, or enteric duplication cyst [22].

b. Although there is a theoretical risk of torsion, current research supports close surveillance and observation of simple ovarian cysts, even those >5 cm, when initially diagnosed prenatally. An estimated 10% of these will regress in utero and roughly half of these will resolve spontaneously in the neonatal period including a third in patients with lesions >5 cm [21, 23].
c. Radiographic heterogeneity, complexity with septae formation or a fluid-debris level are all suggestive of intra-uterine torsion or hemorrhage [21, 22].
d. There are no indications for antenatal intervention or early induction of labor if either an ovarian cyst or ovarian torsion is suspected. Rather, expectant management is recommended with post-natal ultrasound and follow-up surveillance 6–12 weeks following delivery [23].

## Treatment

A. Historical Perspective

   a. Historically, ovarian torsion was treated with oophorectomy, however, recent decades have demonstrated that ovarian preservation is both a feasible and highly effective management strategy.
   b. The various rationales in support of oophorectomy have included (a) leaving a nonviable ovary behind may lead to infection or need for subsequent operation; (b) a malignancy may be missed; (c) patient may sustain a thromboembolic event from the ovarian vessels untwisting during the maneuvering. These theoretical concerns have been subsequently debunked and ovarian sparing surgery is now the preferred surgical intervention for patients with ovarian torsion [1, 2, 5].
   c. Ovarian salvage rates have been reported to be as low as 32% according to a 2016 systematic review with practice pattern differences identified between pediatric general surgeons and gynecologists [3, 12]. A study utilizing the Pediatric Health Inpatient Services (PHIS) database has found that pediatric general surgeons were more likely to perform an oophorectomy than a gynecologist during a similar time period [24]. A 2015 study using the National Inpatient Sample showed that 78% of patients with ovarian torsion had an oophorectomy performed with this occurring more commonly at non-teaching hospitals [25].
   d. Trends over recent decades show an increase in detorsion alone without oophorectomy from 9 to 25% over the study period of 1998 through 2011 [25]. Further demonstrating progress, innumerable recent single-institution studies have confirmed a predilection for ovarian-sparing approaches.

B. Operative Interventions

   a. Laparotomy versus Laparoscopy

      i. Minimally invasive techniques have become increasingly favored in the management of ovarian torsion. Laparoscopy has been shown to decrease length of stay, decrease post-operative complication rate, and result in an improved chance of ovarian preservation [26] (Image 1).
      ii. Laparotomy via a small Pfannenstiel incision has been utilized in cases where torsion occurs in the setting of a large associated mass. This has been shown to facilitate safe delivery of the ovary and associated mass with good cosmesis and rapid recovery post-operatively (Image 2).

   b. Oophorectomy

      i. Oophorectomy should be avoided if possible given evidence that suggests the majority of ovaries thought to be threatened or necrotic intraoperatively are ultimately salvable. Interestingly, a number of studies

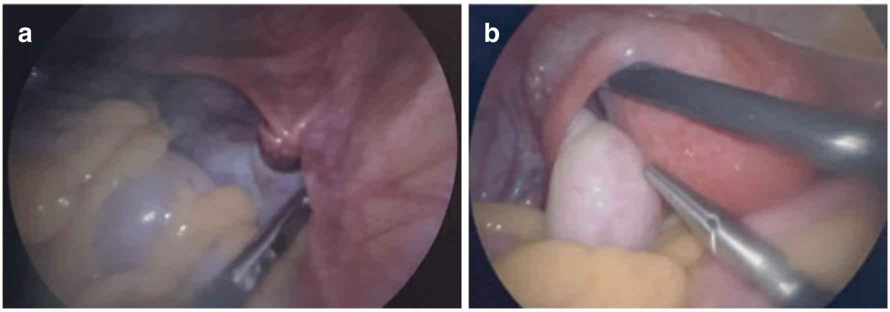

**Image 1** Laparoscopic view of ovarian torsion. (**a**) Right ovarian torsion with twisted adnexal structures, associated enlarged cyst containing ovary and mild-moderate vascular congestion and ischemia. (**b**) Left ovarian torsion with <360° twist and well-perfused slightly edematous ovary. Source: Muriel Cleary, MD

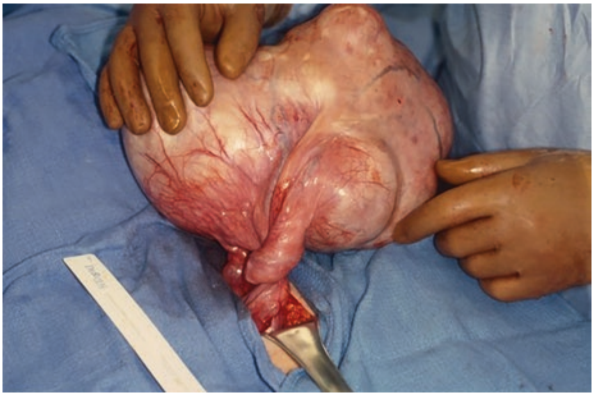

**Image 2** Laparotomy via Pfannenstiel incision for a large right-sided ovarian teratoma presenting with acute torsion. Source: Marc Lessin, MD

    that have prioritized ovarian salvage have shown a 0% rate of re-operation to remove a torsed adnexa [4].
   ii. A 2018 systematic review found that a blue or purplish discolored ovary with presumed necrosis at the time of operative exploration did not correlate with the current or future capacity of an ovary to develop follicles [3]. It was thus their recommendation not to use this gross appearance as the sole determinant of ovarian resection.
   iii. Oophorectomy must be considered if the ovary itself is necrotic beyond salvage and friable such that detorsion alone is not feasible.
   iv. Resection must also be considered in the setting of a confirmed malignancy although the presence of a mass alone has been shown to be falsely concerning with the majority of these ultimately being benign. A large institutional review found an overall malignancy rate of 1.8% similar to findings in other studies looking at rates of a malignant mass after oophorectomy for torsion [27]

c. Detorsion alone

   i. Detorsion without attempted resection of an associated mass has been increasingly proposed in the literature as a viable option in efforts to preserve the ovary.
   ii. Due to the engorgement and edema associated with an obstructed venous outflow, the ovary is often blue-black in appearance, and this does not immediately resolve at the time of detorsion [28].
   iii. Recent studies have demonstrated that after simple detorsion, despite subjective rates of high-grade necrosis, follow-up ultrasounds of the detorsed ovaries demonstrate 100% follicular development [29]. Similar findings have been described in other studies utilizing detorsion alone with follow-up imaging confirming >95% rates of viable functioning ovarian parenchyma [30–32].
   iv. Given the high risk of bleeding complications and damage to ovarian tissue in the setting of an enlarged and edematous ovary, one safe option that maximizes the likelihood of sparing the ovary is to perform detorsion alone [4].
   v. Follow-up imaging and interval laparoscopy with ovarian-preserving surgery can then be performed in the cases in which the ultrasound demonstrates a persistent ovarian mass.

d. Detorsion with Cystectomy

   i. In cases where the ovary appears viable and there is a clearly associated ovarian cyst or mass, another option is to proceed with detorsion and intra-operative ovarian-sparing cystectomy [3, 11].
   ii. This is classically performed in a bi-valving fashion by opening the outer rim of ovarian parenchyma along the longitudinal axis and shelling out the intact cyst (Image 3).

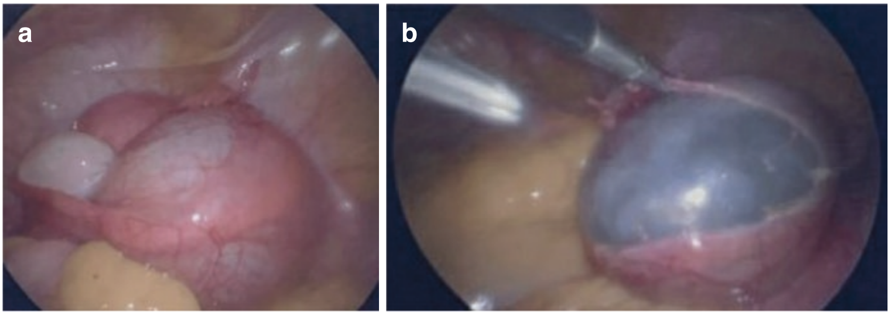

**Image 3** Laparoscopic view of a right-sided ovarian cyst (**a**) Cyst-containing ovary and overlying fallopian tube with minimal edema and no ischemic changes following detorsion of a <360° twist. (**b**) Bi-valving of ovarian parenchyma revealing cyst which was subsequently removed leaving behind viable ovarian tissue. Source: Muriel Cleary, MD

   e. Detorsion with Oophoropexy
      i. There is no convincing evidence to suggest that routine oophoropexy should be performed for patients who undergo detorsion in the absence of an associated cyst or mass.
      ii. Studies, however, have demonstrated recurrent torsion rates between 1.3 and 18% with the majority of these occurring in premenarchal patients without an associated mass [6, 33–35].
      iii. Some studies have proposed oophoropexy by (1) plication of the elongated utero-ovarian ligament and shortening of the adnexal ligaments or by (2) fixation to the pelvic sidewall or uterosacral ligament using nonabsorbable suture [36–38]. With fixation, there is the potential for disruption of normal anatomical configuration with unknown effects on future fertility, therefore, plication appears to be the currently favored approach.
      iv. While this is not done universally as a routine intervention when there is no underlying pathologic mass, oophoropexy should be considered if a patient presents with recurrent torsion.

C. Neonatal Management
   a. Antenatal torsion can become apparent on prenatal ultrasounds that demonstrate complex heterogeneous cysts with layering debris [21, 23, 39]. Immediate post-natal surgical exploration is not routinely recommended if the ovary is deemed already lost as the risks of neonatal surgery exceed any potential benefit that might be conferred. This may ultimately result in autoamputation and can sometimes manifest in a “wandering ovary” [39] . These ovaries may require operative intervention beyond the immediate neonatal period, ideally via laparoscopy, but can also be managed expectantly [23, 39].

  b. It has been estimated that 16% of pediatric ovarian torsions occur in the first year of life and presenting symptoms can be more challenging to detect given that the patient is non-verbal [27].
  c. One recent institutional study found that overall, only 4% of patients with a neonatal cyst undergoing surveillance developed torsion and only 13% of those with cysts >5 cm developed torsion [23].
  d. A similarly low threshold for diagnostic laparoscopy should be maintained if acute ovarian torsion is suspected. However, not all neonatal ovarian cysts need to be urgently explored as a period of observation does not decrease the patient's chances of ovarian salvage and may prevent upwards of 50% of neonates from undergoing general anesthesia [3, 23].

## Outcomes

A. Risk of Re-torsion

  a. The risk of recurrent ovarian torsion is thought to be more common in premenarchal patients with normal ovaries and may be due to laxity of the utero-ovarian ligaments combined with hormonal hyperstimulation and the resultant increase in ovarian size that occurs during this time [7, 35].
  b. In one study of pediatric patients, a recurrence rate of 3.4% was identified with none of the patients having an associated pathologic lesion at the initial operation [4].
  c. For patients with recurrent torsion without an associated adnexal mass, oophoropexy has been suggested as an effective intervention at the time of re-operation [40].

B. Rate of associated malignancy

  a. It can be challenging to determine if there is an associated mass in cases of ovarian torsion where the ovary is edematous and congested.
  b. In the adult ovarian torsion population, the incidence of a concurrent malignancy is 2% [18]. To examine this risk in the pediatric population, a systematic review conducted by the American Pediatric Surgical Association Outcomes and Evidence-Based Practice Committee showed that malignancies were found in only 0.4–5% of resected ovaries and determined that there is no evidence to support a benefit of oophorectomy over detorsion for fear of missing an associated malignancy [3].
  c. A large single-institution study suggested that among a pediatric population with an occult malignancy rate of 1.8%, there was a greater risk of associated malignancy for those with ovarian masses >8 cm [27].

C. Surveillance post-operative imaging

  a. Post-operative ultrasound is useful to document the return of normal ovarian size, to follow up the presence of an associated mass, and to demonstrate normal ovarian follicles.
  b. One study noted that it may take up to 6 months for normal blood flow to be restored after detorsion however the majority of follicular development will occur by 3 months [41].
  c. Most sources recommend follow-up ultrasound at 3 months after ovarian cystectomy or detorsion [2, 3, 18].

D. Future Fertility

  a. Data examining future fertility in patients who have undergone surgery for ovarian torsion is limited.
  b. Studies looking at the presence of follicles in detorsed ovaries have demonstrated that >88–100% of ovaries have follicles on follow-up ultrasound [2, 30].
  c. Long-term studies have also looked at rates of successful pregnancies with several case series reporting successful pregnancies and live births in patients who had undergone ovarian detorsion in the past [2, 3].
  d. In one report, in vitro fertilization was necessary in only 5% of patients with a history of ovarian torsion who had difficulty becoming pregnant [42].

## Additional Notes

A. Predictive scoring tools used to guide operative intervention:

Ovarian torsion has a varied clinical presentation such that a scoring system would be valuable to aid in making the diagnosis. Currently, there are no validated widely accepted scoring systems that are commonly used to direct surgical decision-making. Some studies have demonstrated the presence of nausea/vomiting, leukocytosis, ovarian volume and ratio as significant composite factors capable of predicting >90% of patients with confirmed torsion [43]. Another study found four factors that were independently associated with ovarian torsion: ovarian ratio, ovarian volume, nausea, and duration of pain. This composite index notably did not include arterial and venous Doppler flows. It found no cases of torsion in patients with a score $< 3$ whereas a score $\geq 3$ had 100% sensitivity and 65.3% specificity and a score $\geq 5$ had 100% specificity. Some groups have explored developing a machine-learned algorithm using sonographic features to guide the decision tree. One such study found that no single variable was sufficient to make a strong prediction however combining information regarding intra-ovarian flow, peripheral follicles, the volume of both ovaries and the presence of an ovarian mass or cyst yielded an AUC of 0.96 ± 0.07 [44]. It remains a clinical and ultimately surgical diagnosis and the threshold to perform a diagnostic laparoscopy should remain low as ovarian salvage depends upon the speed with which the vessels can be detorsed.

B. Thresholds to perform detorsion without cystectomy:

In the setting of an acutely engorged and ischemic ovary, it can be challenging both radiographically and operatively to determine whether there is a true discrete ovarian mass involved. Additionally, even if there is a confirmed suspicious or complex mass, current data suggests that the risk of it being malignant is statistically quite low [27]. Studies urge simple detorsion if the ovary is edematous and friable making cystectomy a higher risk procedure with an increased likelihood of damaging potentially salvable ovarian tissue. Current position statements from the American College of Obstetricians and Gynecologists and the American Pediatric Surgical Association support a similar approach unless a pre-operative work-up has confirmed a malignancy [3, 11]. Outcomes show that ovaries that have undergone detorsion alone normalize in size and demonstrate normal follicular development on follow-up ultrasound in 93–100% of cases [28, 31, 42]. For those with concerning features, tumor markers may subsequently be sent and additional imaging obtained to optimize the surgical plan and favor an ovarian-sparing approach where appropriate.

## Questions

1. A 1-day old asymptomatic baby girl with a prenatal diagnosis of a 3-cm intrabdominal cyst presents with a post-natal ultrasound that reveals a 3.5-cm simple cyst that appears to be associated with the right ovary. The left ovary is not well seen. What is the next best step?

(A) Diagnostic laparoscopy with ovarian cystectomy
(B) Diagnostic laparotomy with ovarian cystectomy
(C) Abdominal CT or MRI
(D) Surveillance ultrasound in 6 weeks

Answer to Question 1: (d) An asymptomatic neonate with a prenatal diagnosis of a simple ovarian cyst is likely to experience spontaneous regression of the cyst and not require intervention. In the absence of clinical or radiographic findings concerning acute torsion, the appropriate management would be with surveillance with post-natal ultrasound in 6–12 weeks. Urgent intervention with laparoscopic or laparotomy is not indicated. Additional imaging is not necessary unless concerning features were seen on ultrasound. Abdominal CT is generally avoided unless absolutely necessary in neonates given associated radiation risks.

2. A 9-year-old girl presents with less than 1 day of acute onset intermittent right lower quadrant abdominal pain and nausea. Her appendix is normal on ultrasound. Which radiographic finding would prompt surgical exploration?

   (A) Pelvic free fluid
   (B) An enlarged right ovary relative to the left (volume of 60 ml versus 4 ml)
   (C) A 3-cm right-sided hemorrhagic cyst
   (D) Normal arterial and venous waveforms on Doppler

Answer to Question 2: (b) Asymmetry of the ovaries, particularly ratios >12–15, have been shown to be strongly associated with an underlying torsion. This results from compression of the twisted ovary's venous outflow and the resulting edema and engorgement. Pelvic-free fluid is a relatively non-specific finding and may be identified in a variety of clinical settings. A hemorrhagic luteal cyst is often 2.5–3.5 in size and can cause pain in the absence of torsion. Normal arterial and venous flow on Doppler does not rule out torsion however would not necessarily prompt one to surgically explore in the absence of other findings.

3. An 11-year-old girl presents with acute onset abdominal pain and is found on ultrasound to have a large 12 cm mixed cystic solid mass associated with a clear right-sided ovarian torsion. Assuming the ovary was edematous and severely ischemic, what would be your next step?

   (A) Obtain an MRI in the next 6–8 h
   (B) Laparoscopy with oophorectomy
   (C) Laparotomy via Pfannenstiel with oophorectomy
   (D) Laparoscopy with detorsion alone and send tumor markers

Answer to Question 3: (d) If the ovary was found to be significant ischemic (blue-black and friable), simple laparoscopic detorsion would be the favored next step. Tumor markers and more detailed cross-sectional imaging can then be obtained followed by definitive interval surgery. The acuity of the torsion precludes waiting for an MRI in this setting. Laparoscopy for a >10–12 cm mass can be higher risk given the potential associated malignancy and risk of spillage. Oophorectomy without confirmation of an underlying malignancy is not optimal management and ovarian-sparing surgery should be pursued if possible.

## References

1. Guthrie BD, Adler MD, Powell EC. Incidence and trends of pediatric ovarian torsion hospitalizations in the United States, 2000-2006. Pediatrics. 2010;125(3):532.
2. Nassif J, Khoury S, Mouanness M. Ovary-sparing surgery for ovarian torsion in adolescents. In: Endometriosis in adolescents. Cham: Springer International; 2020. p. 511–27.
3. Dasgupta R, Renaud E, Goldin AB, et al. Ovarian torsion in pediatric and adolescent patients: a systematic review. J Pediatr Surg. 2018;53(7):1387–91.

4. Adeyemi-Fowode O, Lin EG, Syed F, Sangi-Haghpeykar H, Zhu H, Dietrich JE. Adnexal torsion in children and adolescents: a retrospective review of 245 cases at a single Institution. J Pediatr Adolesc Gynecol. 2019;32(1):64–9.
5. Oltmann SC, Fischer A, Barber R, Huang R, Hicks B, Garcia N. Cannot exclude torsion—a 15-year review. J Pediatr Surg. 2009;44(6):1212–6. discussion 1217
6. Lawrence AE, Minneci PC, Deans KJ. Ovarian masses and torsion: new approaches for ovarian salvage. Adv Pediatr. 2020;67:113–21.
7. Nanni M, Merola MG, Ianniello S, Orazi C, Schingo PM, Trinci M. Ovarian torsion. In: Imaging non-traumatic abdominal emergencies in pediatric patients. Cham: Springer International; 2016. p. 205–27.
8. Ito K, Utano K, Kanazawa H, et al. CT prediction of the degree of ovarian torsion. Jpn J Radiol. 2015;33(8):487–93.
9. Tsafrir Z, Azem F, Hasson J, et al. Risk factors, symptoms, and treatment of ovarian torsion in children: the twelve-year experience of one center. J Minim Invasive Gynecol. 2012;19(1):29–33.
10. Servaes S, Zurakowski D, Laufer MR, Feins N, Chow JS. Sonographic findings of ovarian torsion in children. Pediatr Radiol. 2007;37(5):446–51.
11. Adnexal torsion in adolescents. ACOG Committee Opinion No, 783. Obstet Gynecol. 2019;134(2):e56–63.
12. Rey-Bellet Gasser C, Gehri M, Joseph JM, Pauchard JY. Is it ovarian torsion? A systematic literature review and evaluation of prediction signs. Pediatr Emerg Care. 2016;32(4):256–61.
13. Kives S, Gascon S, Dubuc É, van Eyk N, No. 341-Diagnosis and management of adnexal torsion in children, adolescents, and adults. J Obstet Gynaecol Can. 2017;39(2):82–90.
14. Jain KA. Sonographic spectrum of hemorrhagic ovarian cysts. J Ultrasound Med. 2002;21(8):879–86.
15. Bronstein ME, Pandya S, Snyder CW, Shi Q, Muensterer OJ. A meta-analysis of B-mode ultrasound, Doppler ultrasound, and computed tomography to diagnose pediatric ovarian torsion. Eur J Pediatr Surg Zeitschrift Kinderchirurgie. 2015;25(1):82–6.
16. Wilkinson C, Sanderson A. Adnexal torsion—a multimodality imaging review. Clin Radiol. 2012;67(5):476–83.
17. Lee EJ, Kwon HC, Joo HJ, Suh JH, Fleischer AC. Diagnosis of ovarian torsion with color Doppler sonography: depiction of twisted vascular pedicle. J Ultrasound Med. 1998;17(2):83–9.
18. Sasaki KJ, Miller CE. Adnexal torsion: review of the literature. J Minim Invasive Gynecol. 2014;21(2):196–202.
19. Peña JE, Ufberg D, Cooney N, Denis AL. Usefulness of Doppler sonography in the diagnosis of ovarian torsion. Fertil Steril. 2000;73(5):1047–50.
20. Chiou SY, Lev-Toaff AS, Masuda E, Feld RI, Bergin D. Adnexal torsion: new clinical and imaging observations by sonography, computed tomography, and magnetic resonance imaging. J Ultrasound Med. 2007;26(10):1289–301.
21. Akalin M, Demirci O, Dayan E, Odacilar AS, Ocal A, Celayir A. Natural history of fetal ovarian cysts in the prenatal and postnatal periods. J Clin Ultrasound. 2021;49(8):822–7.
22. di Giacomo V, Trinci M, van der Byl G, Catania VD, Calisti A, Miele V. Ultrasound in newborns and children suffering from non-traumatic acute abdominal pain: imaging with clinical and surgical correlation. J Ultrasound. 2015;18(4):385–93.
23. Papic JC, Billmire DF, Rescorla FJ, Finnell SME, Leys CM. Management of neonatal ovarian cysts and its effect on ovarian preservation. J Pediatr Surg. 2014;49(6):990–3. discussion 993-4
24. Campbell BT, Austin DM, Kahn O, et al. Current trends in the surgical treatment of pediatric ovarian torsion: we can do better. J Pediatr Surg. 2015;50(8):1374–7.
25. Sola R, Wormer BA, Walters AL, Heniford BT, Schulman AM. National trends in the surgical treatment of ovarian torsion in children: an analysis of 2041 pediatric patients utilizing the nationwide inpatient sample. Am Surg. 2015;81(9):844–8.

26. Lo LM, Chang SD, Horng SG, Yang TY, Lee CL, Liang CC. Laparoscopy versus laparotomy for surgical intervention of ovarian torsion. J Obstet Gynaecol Res. 2008;34(6):1020–5.
27. Oltmann SC, Fischer A, Barber R, Huang R, Hicks B, Garcia N. Pediatric ovarian malignancy presenting as ovarian torsion: incidence and relevance. J Pediatr Surg. 2010;45(1):135–9.
28. Cohen SB, Wattiez A, Seidman DS, et al. Laparoscopy versus laparotomy for detorsion and sparing of twisted ischemic adnexa. JSLS. 2003;7(4):295–9.
29. Parelkar SV, Mundada D, Sanghvi BV, et al. Should the ovary always be conserved in torsion? A tertiary care institute experience. J Pediatr Surg. 2014;49(3):465–8.
30. Geimanaite L, Trainavicius K. Pediatric ovarian torsion: follow- up after preservation of ovarian tissue. J Pediatr Surg. 2019;54(7):1453–6.
31. Santos XM, Cass DL, Dietrich JE. Outcome following detorsion of torsed adnexa in children. J Pediatr Adolesc Gynecol. 2015;28(3):136–8.
32. Aziz D, Davis V, Allen L, Langer JC. Ovarian torsion in children: is oophorectomy necessary? J Pediatr Surg. 2004;39(5):750–3.
33. Beaunoyer M, Chapdelaine J, Bouchard S, Ouimet A. Asynchronous bilateral ovarian torsion. J Pediatr Surg. 2004;39(5):746–9.
34. Melcer Y, Sarig-Meth T, Maymon R, Pansky M, Vaknin Z, Smorgick N. Similar but different: a comparison of adnexal torsion in pediatric, adolescent, and pregnant and Reproductive-Age Women. J Women's Health. 2016;25(4):391–6.
35. Smorgick N, Melcer Y, Sarig-Meth T, Maymon R, Vaknin Z, Pansky M. High risk of recurrent torsion in premenarchal girls with torsion of normal adnexa. Fertil Steril. 2016;105(6):1561–1565.e3.
36. Crouch NS, Gyampoh B, Cutner AS, Creighton SM. Ovarian torsion: to pex or not to pex? Case report and review of the literature. J Pediatr Adolesc Gynecol. 2003;16(6):381–4.
37. Comeau IM, Hubner N, Kives SL, Allen LM. Rates and technique for oophoropexy in pediatric ovarian torsion: A single-institution case series. J Pediatr Adolesc Gynecol. 2017;30(3):418–21.
38. Fuchs N, Smorgick N, Tovbin Y, et al. Oophoropexy to prevent adnexal torsion: how, when, and for whom? J Minim Invasive Gynecol. 2010;17(2):205–8.
39. Ozcan HN, Balci S, Ekinci S, et al. Imaging findings of fetal-neonatal ovarian cysts complicated with ovarian torsion and autoamputation. AJR Am J Roentgenol. 2015;205(1):185–9.
40. Germain M, Rarick T, Robins E. Management of intermittent ovarian torsion by laparoscopic oophoropexy. Obstet Gynecol. 1996;88(4 Pt 2):715–7.
41. Templeman C, Hertweck SP, Fallat ME. The clinical course of unresected ovarian torsion. J Pediatr Surg. 2000;35(9):1385–7.
42. Oelsner G, Bider D, Goldenberg M, Admon D, Mashiach S. Long-term follow-up of the twisted ischemic adnexa managed by detorsion. Fertil Steril. 1993;60(6):976–9.
43. Umasankar N, Howard SM, Tomlin KV. Adnexal torsion predictive tool maintains applicability in a heterogeneous pediatric population. J Pediatr Adolesc Gynecol. 2021;34(5):656–61.
44. Otjen JP, Stanescu AL, Alessio AM, Parisi MT. Ovarian torsion: developing a machine-learned algorithm for diagnosis. Pediatr Radiol. 2020;50(5):706–14.

# Testicular Masses

**Jasmine I. Kashkoush and Robert Steckler**

**Key Points**

1) Men with cryptorchidism are four to six times more likely to be diagnosed with testis cancer. The relative risk decreases if orchidopexy is performed prior to puberty.
2) In patients with a testis mass or unexplained scrotal signs, a scrotal ultrasonography should be considered as an extension of the physical examination. However, ultrasonography cannot distinguish between malignant and benign lesions.
3) Pediatric testicular masses are generally benign, whereas adult testicular tumors are malignant.

Pediatric testicular masses differ significantly from those in the adult population. Prepubertal testicular tumors are more commonly benign and those that are malignant have a more favorable course.

1. Epidemiology
    a. Most common diagnosis in infants is a yolk sac tumor.
    b. Generally, will see around 2 years old or peripubertal

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2. Pathogenesis
   a. Cryptorchidism is a risk factor. Early orchidopexy may mitigate the increased risk of malignancy to baseline.
   b. Patients with disorders of sexual differentiation are at increased risk of malignancy if they have Y material.
3. Clinical Presentation
   a. Painless testicular mass
   b. Incidentally as a lesion on scrotal ultrasound
4. Diagnosis
   a. Can obtain a scrotal ultrasound. These are very sensitive.
      i. However, do not distinguish benign from malignant lesions.
      ii. Teratoma: may appear heterogenous and complex.
      iii. Yolk sac tumor: well-circumscribed and heterogenous.
      iv. Mixed germ cell tumors.
      v. Epidermoid cysts: typically look like "onion skin" made of alternation hypo and hyperechoic lesions.
   b. Biomarkers
      i. AFP
         1. Normally produced by fetal yolk sac, liver, and GI tract.
         2. Elevated in yolk sac tumor.
         3. Will be physiologically elevated in children <1 years old.
         4. Not secreted by seminoma or choriocarcinoma.
         5. Half-life is 5–7 days.
      ii. Beta1-HCG
         1. Rarely elevated in prepubertal testicular tumors.
         2. Secreted by syncytiotrophoblasts.
         3. May be found in non-seminomatous germ cell tumors and in seminomas.
         4. Half-life is 24–36 h.
      iii. LDH
         1. Half-life is 24 h
         2. Not specific for germ cell tumors, but reflects high tumor burden
5. Tumor Type
   a. Germ cell tumors
      i. Teratoma

            1. Combination of three primitive embryologic germ cell layers
            2. Do not express AFP
            3. Treatment: partial orchiectomy if prepubertal
        - ii. Yolk sac tumor
            1. Most common malignant prepubertal tumor
            2. Schiller-Duval bodies are pathognomonic
            3. Elevated AFP
    - b. Gonadal Stromal Tumors
        - i. Leydig cell tumor
            1. Hormonally active, associated with precocious puberty
            2. May be small and appear as yellow-brown nodules
            3. Histology: Diffuse layers of polygonal cells and 40% have Reinke crystals
            4. May have elevated testosterone
            5. Recurrence not common
        - ii. Sertoli cell tumors
            1. Present earlier than Leydig cells
            2. Associated with Peutz-Jeghers and Carney syndromes
            3. Firm, well-circumscribed, and tan-gray appearance with areas of hemorrhage or cysts
            4. Treatment: <5 years old may do orchiectomy. If >5 years old and with >5 cm, vascular invasion, necrosis, cellular atypia needs to undergo full staging workup
    - c. Leukemia and Lymphoma
        - i. Most common cancers metastatic to the testis.
        - ii. High success rates with systemic chemotherapy and whole-body irradiation. Do not need to perform routine testis biopsy.

## Additional Notes

Serum biomarkers are an important role when diagnosing and evaluating testicular masses in both the pediatric and adult populations. B-HCG is rarely elevated in pediatric tumors, whereas AFP is commonly elevated.

The role of AFP must be evaluated carefully. AFP is normally produced by the fetal yolk sac, liver, and gastrointestinal tract. It is also physiologically elevated in children up to 1 year of age. Thus, the interpretation of AFP must be done with caution. AFP is elevated in yolk sac tumor. If a child is older than 1 year of age with an elevated AFP, this is likely to be secondary to a yolk sac tumor.

Although most pediatric prepubertal testicular masses are benign and can be managed with organ-sparing procedures, a yolk sac tumor should not. A yolk sac tumor is the most common prepubertal malignant lesion. The schiller-Duval bodies are pathognomonic for yolk sac tumor.

## Study Questions

1) Question 1: A 5-year-old male presents to the clinic with a painless testicular mass. His scrotal ultrasound confirms a right testicular mass. Serum biomarkers reveal an AFP of 100. What is the likely diagnosis?
   a. Yolk sac tumor
   b. Seminoma
   c. Choriocarcinoma
   d. Teratoma

Answer to Question 1: (a.) Yolk sac tumor. Patients <1 year of age have normal physiologic elevations of their AFP. In patients >1 year of age who present with a testicular mass and an elevated AFP, they likely have a yolk sac tumor. Pure choriocarcinoma, teratoma, and seminoma do not secrete AFP.

2) Question 2: Which of the follow are risk factors for testicular cancer?
   a. White race
   b. History of cryptorchidism
   c. Family history of testicular cancer
   d. All of the above.

Answer to Question 2: (d.) All of the above. These are all risk factors for the development of testicular cancer. All of these factors increase the risk of testicular cancer.

3) Question 3: Which organ normally produces AFP?
   a. Liver
   b. Brain
   c. Testis
   d. Heart

Answer to Question 3: (a.) Liver. AFP is normally produced by the liver, yolk sac, and gastrointestinal systems. There is physiologic elevation up until 1 year of age.

## Further Reading

AUA University. Update series (2019) lesson 38: testicular tumors in children and adolescents. Linthicum, MD: American Urological Association. 2019. Online. https://auau.auanet.org/content/course-318. Accessed 9 Nov 2021.

Partin AW, Dmochowski RR, Kavoussi LR, Peters CA, Wein AJ. Campbell walsh wein urology. 12th ed. Amsterdam, NL: Elsevier; 2020.

Smith JA, Howards SS, Preminger GM, Dmochowski RR. Hinman's atlas of urologic surgery revised reprint. 4th ed. Amsterdam, NL: Elsevier; 2019.

# Adrenal Tumors

**Rebekah Campbell and Melissa Mao**

**Key Points**

1) There are several syndromes that are associated with adrenal tumors, and these often need to be worked up prior to management of the adrenal tumor.
2) Incidentalomas are increasingly more common with the wider use of cross-sectional imaging. A functional workup should be undertaken in all incidentalomas.
3) Biopsies are rarely indicated and should not be done prior to ruling out a pheochromocytoma.
4) Surgical treatment can be curative for benign tumors.

## I. Background

### A. Embryology:

1. The cortex arises from the mesoderm, and the medulla from the neuroectoderm.
2. Cortex starts to develop in 5th week gestation, and is functional at 7 weeks.
3. Fetus produces large amounts of Dehydroepiandrosterone (DHEA-S) and this ultimately affects estrogen production in the mother. Estrogen increases DHEA-S creating a positive feedback loop.

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4. Catecholamines are released in response to hypoxia and hypoglycemia at 5 months gestation.
5. At the end of gestation, the gland consists of glomerulosa, transitory, and fetal zones. The fetal zone involutes until 2 years of life.

B. Anatomy/Physiology

1. Location: There are two adrenal glands, superior to the kidneys.
2. Arterial supply is from aorta, renal, and phrenic arteries.
3. Venous drainage: The left adrenal vein drains into the left renal vein. The right adrenal vein drains directly into the inferior vena cava.
4. Gross Description: Adrenal gland is larger in neonates and infants, and regresses with age. They are generally golden in color.
5. Histology: Adrenal gland consists of outer cortex and inner medulla
    a. Cortex: glomerulosa secretes aldosterone, fasciculata secretes cortisol, and reticularis secretes androgens. The reticularis starts to develop at 4–8 years old and reaches maturity at 10–20 years old.
    b. Medulla: secretes catecholamines.

C. Adrenal pathology

1. Congenital Adrenal Hyperplasia: group of autosomal recessive disorders. Most common form is due to 21-hydroxylase deficiency. Range of symptoms including virilization, hyponatremia, hyperkalemia, hypotension. Can present in infancy to adults.
2. Adrenal Cysts
3. Tumors
    a. Benign non-functioning adenoma.
    b. Functional Tumors: Aldosteronomas, Cushing's syndrome, and Pheochromocytomas.
    c. Neuroblastoma: one of the most common pediatric malignancies.
    d. Adrenocortical carcinoma.
    e. Metastasis from another primary cancer.
4. Hemorrhage/Trauma
    a. Hemorrhage is the most common cause of adrenal mass in neonates.

D. Association with syndromes:

1. Pheochromocytoma: Multiple Endocrine Neoplasia Type 2, Neurofibromatosis Type 1, Von Hippel–Lindau Syndrome, Paraganglioma-Pheochromocytoma Syndromes, Tuberous Sclerosis, Sturge–Weber Syndrome, Carney's Triad.
2. Adrenocortical Carcinoma: Li–Fraumeni Syndrome, Beckwith–Wiedemann, Familial Adenomatous Polyposis, Multiple Endocrine Neoplasia Type 1.

3. Neuroblastoma: Li–Fraumeni Syndrome, Beckwith–Wiedemann, Paraganglioma-Pheochromocytoma Syndromes, Weaver Syndrome, Fanconi Anemia.

II. Diagnosis

A. History: Important to inquire about family history. Should also ask about blood pressure issues, palpitations, diaphoresis, headaches, muscle cramping, hypokalemia, hyperglycemia, virilization/feminization, and abdominal discomfort.
B. Physical exam: Check blood pressure, perform respiratory, cardiac, and abdominal exams, and evaluate for skin changes/morphology associated with Cushing's Syndrome.
C. Serum/Urine studies: Rule out functional mass.

   1. Cushing's Syndrome: 1 mg dexamethasone suppression test, 24-h urine cortisol, midnight salivary cortisol.
   2. Hyperaldosteronism: Basic metabolic panel, Plasma aldosterone:renin ratio, Saline suppression test.
   3. Pheochromocytoma: Plasma and 24-h urine metanephrines/normetanephrines.
   4. Sex hormones: Serum testosterone, DHEA-S, Estradiol.
   5. Adrenocortical carcinoma: commonly hypersecretes sex hormones (virilization) ± cortisol (Cushing's Syndrome).
   6. Neuroblastoma: Urine homovanillic acid (HVA) and vanillylmandelic acid (VMA).
   7. Congenital Adrenal Hyperplasia: electrolytes, plasma renin, 17-hydroxyprogesterone, androstenedione, and progesterone.

D. Imaging

   1. Ultrasound: most useful in prenatal and neonatal periods.
   2. Magnetic Resonance Imaging (MRI)

      a. Recommended for surveillance in suspected benign lesions.
      b. May require sedation.

   3. Computed Tomography (CT)

      a. Recommended for preoperative evaluation.
      b. Concern for radiation exposure, use single-phase CT.

   4. Meta-iodobenzylguanidine (MIBG) scan

      a. Nuclear medicine scan.
      b. Useful for detecting pheochromocytoma, neuroblastoma.

   5. Positron Emission Tomography (PET)

      a. Fluorodeoxyglucose (FDG) and dotatate PET scans: both options to assess for metastasis/malignant potential.

III. Treatment

A. Surgical

1. Indications
    a. Hormonally active tumor.
    b. Enlarging adrenal incidentaloma.
    c. Adrenal incidentalomas with size $\geq$ 4 cm.
    d. Concern for malignancy, metastatic disease, or neuroblastoma
        i. Biopsy may be needed for diagnosis.
        ii. Must first rule out pheochromocytoma prior to biopsy.
2. Preoperative considerations
    a. Pheochromocytoma: alpha blockade; good communication with anesthesia, arterial line intraoperatively.
    b. Cushing's Syndrome: plan for steroids postoperatively due to reflexive adrenal insufficiency.
3. Approach
    a. Laparoscopic vs. open transperitoneal: Laparoscopic is becoming more preferred, open is reserved for large tumors and malignancy
        i. Right adrenalectomy: retract liver, shorter adrenal vein directly into the inferior vena cava.
        ii. Left adrenalectomy: Retract spleen, may need to mobilize splenic flexure of colon, be aware of tail of pancreas and splenic artery.
    b. Laparoscopic posterior retroperitoneal: best for small tumors, history of prior abdominal surgery with concern for adhesions.
    c. Cortical-sparing/partial adrenalectomy: useful in patients with bilateral tumors in order to avoid need for post-operative corticosteroids. Use intraoperative ultrasound to guide resection.

B. Nonoperative: Interval imaging in 3 months. If no change or growth, consider resection.

C. Post-operative considerations

1. Intraoperative complications: Damage to nearby structures (pancreas, spleen, splenic artery, colon on the left and duodenum, inferior vena cava on the right).
2. Adrenal insufficiency: expect after removing a tumor causing Cushing's Syndrome (transient, treat with glucocorticoids). For bilateral adrenalectomy patients, treat with gluco- and mineralo-corticoids.
3. Surgery can be curative for benign conditions.
4. Neuroblastoma has a survival rate that ranges from 98% in low-risk groups, and 40–50% in high-risk groups. Moderate- and high-risk groups (based on biologic characteristics) should be monitored for recurrence.

5. Pheochromocytoma has an excellent survival and recurrence rate in benign disease. For malignant pheochromocytomas (local invasion, metastasis) the 5-year survival is 78%, and 15-year survival is 31%. Recurrence can be after many years, and so long-term follow-up is necessary.
6. Adrenocortical carcinoma has a poor prognosis (54% 5-year survival).
7. Post-operative monitoring for hormonally active tumors should consist of hormonal testing every 2 months for the first year, every 4 months for the second year, and every 6 months beyond this.
8. Yearly CT or MRI for 5 years after resection.

D. Chemotherapy may be used for locally advanced/recurrent adrenocortical carcinomas and neuroblastoma.
E. Treatment of congenital adrenal hyperplasia may include mineralocorticoid and glucocorticoid replacement, and ensuring normal puberty and development.

## Additional Notes

The adrenal glands were first described by an Italian anatomist Bartolomeo Eustachius in 1563, who called them "glandulae renibus incumbents." However, the function of the adrenal glands was not discovered until the mid-1800s. Key discoveries were made by Thomas Addison in 1853, who described the sequelae of adrenal insufficiency (Addison's disease); Harvey Cushing in 1932, who described the manifestations of pituitary abnormalities resulting in excess secretion from the adrenals (Cushing's disease); and Jerome Conn in 1955, who described primary aldosteronism (Conn's disease). Charles Mayo performed the first adrenalectomy in the USA in 1927 for a pheochromocytoma.

Of all pediatric adrenal tumors, neuroblastomas account for over 90%. Derived from the adrenal medulla, neuroblastomas represent 8–10% of all childhood tumors, and are the most common extra-cranial solid tumor in the pediatric population. They account for 15% of all pediatric cancer-related deaths. In the USA, approximately 800 cases are diagnosed each year, with most cases diagnosed before age 5. In contrast, pediatric tumors of the adrenal cortex are very rare. Adrenocortical carcinoma represents approximately 0.2% of childhood malignancies, which translates to about 25 cases diagnosed annually in the USA.

While laparoscopic adrenalectomy has become the standard surgical approach in adults, only recently has it become adopted as a preferred approach in children. There has been growing evidence to support the use of laparoscopic adrenalectomy in children, demonstrating both good outcomes and few complications. The most commonly used approach is the laparoscopic transperitoneal approach. Other less commonly used approaches include laparoscopic posterior retroperitoneal, single site, and robotic.

## Study Questions

1) Which familial syndromes are associated with adrenal tumors?

   a. Medullary Endocrine Neoplasia type 1, Beckwith–Wiedemann Syndrome, and Neurofibromatosis type 1.
   b. Von–Hippel Lindau Syndrome, Lynch Syndrome, and Li–Fraumeni Syndrome.
   c. Cowden Syndrome, Medullary Endocrine Neoplasia type 2a, and Familial Adenomatous Polyposis Syndrome.
   d. Li–Fraumeni Syndrome, Paraganglioma-Pheochromocytoma Syndrome, and Cowden Syndrome.

Answer to Question 1: (a.) Medullary Endocrine Neoplasia type 1, Beckwith-Wiedemann Syndrome, and Neurofibromatosis type 1.

Medullary Endocrine Neoplasia type 1 is associated with adrenocortical carcinoma, and Medullary Endocrine Neoplasia type 2a and 2b are associated with pheochromocytomas. Beckwith–Wiedemann Syndrome and Li–Fraumeni Syndrome are associated with adrenocortical carcinoma and neuroblastoma. Neurofibromatosis type 1 and Von–Hippel Lindau are associated with pheochromocytoma. Paraganglioma-Pheochromocytoma syndrome is associated with pheochromocytoma and neuroblastoma. Familial Adenomatous Polyposis syndrome is associated with adrenocortical carcinoma. Lynch Syndrome and Cowden Syndrome are not associated with adrenal tumors.

2) A 14-year-old male was involved in a motor vehicle accident 1 month ago, and is fully recovered. CT scan obtained at the time of the accident shows a 0.5 cm right adrenal nodule. Which of the following would not be included in his initial workup?

   a. DHEA-Sulfate
   b. 24-h urine metanephrines
   c. MIBG Scan
   d. Aldosterone

Answer to Question 2: (c.) MIBG Scan

A biochemical workup should always be obtained for any incidentaloma. This consists of looking for aldosteronoma by checking aldosterone-to-renin ratio, for Cushing's syndrome by checking cortisol, for DHEA-Sulfate (overexpression generally is seen with adrenocortical carcinoma), and for pheochromocytoma by checking for metanephrines. The following should be obtained: aldosterone, renin, either 24-h urine cortisol or low-dose suppression test, DHEA-S, and either plasma metanephrines or 24-h urine metanephrines. MIBG scan is used to localize the source of epinephrine/norepinephrine oversecretion. This is not obtained during the initial evaluation.

3) When is a biopsy of the adrenal gland appropriate?
    a. When there is concern for malignancy after obtaining cross-sectional imaging
    b. When there is a >4 cm adrenal incidentaloma
    c. When there is a known primary cancer, and there is concern for metastatic disease
    d. When there is concern for malignancy, and a pheochromocytoma has been ruled out

Answer to Question 3: (d.) When there is concern for malignancy, and a pheochromocytoma has been ruled out

A biopsy of the adrenal gland should never be done unless a pheochromocytoma has been ruled out with biochemical testing—either 24-h urine metanephrines, or plasma metanephrines. The concern for primary adrenal malignancy or metastatic disease is an indication for biopsy, but only after metanephrines are found to be normal.

## Further Reading

Alexander F. Neuroblastoma. Urol Clin N Am. 2000;27(3):383–92.

Barr EK, Applebaum MA. Genetic predisposition to neuroblastoma. Children (Basel). 2018;5(9):119.

Bholah R, Bunchman TE. Review of pediatric pheochromocytoma and paraganglioma. Front Pediatr. 2017;5:155.

Colon NC, Chung DH. Neuroblastoma, advanced. Pediatrics. 2011;58(1):297–311.

Lee YT, Samsudin H, Ong CCP, et al. Posterior retroperitoneoscopic adrenalectomy for pediatric adrenal tumors. J Pediatr Surg. 2019;54(11):2348–52.

Maris JM. Recent advances in neuroblastoma. N Engl J Med. 2010;362(23):2202–11.

Papadakis M, Manios A, Schoretsanitis G, et al. Landmarks in the history of adrenal surgery. Hormones. 2015;15(1):136–41.

Sutter JA, Grimberg A. Adrenocortical tumors and hyperplasias in childhood—etiology, genetics, clinical presentation and therapy. Pediatr Endocrinol Rev. 2006;4(1):32–9.

Wengler CA, Nolan HR, Glenn J. Laparoscopic adrenalectomy in children. In: Walsh D, Ponsky T, Bruns N, editors. The SAGES manual of pediatric minimally invasive surgery. Cham: Springer; 2017.

Witchel SF. Congenital adrenal hyperplasia. J Pediatr Adolesc Gynecol. 2017;30(5):520–34.

Xu X, Sergi C. Pediatric adrenal cortical carcinomas: histopathological criteria and clinical trials. A systematic review. Contemp Clin Trials. 2016;50:37–44.

# Pancreatitis

**Elizabeth J. Hughes and Marybeth Browne**

**Key Points**

1. Pancreatitis is inflammation of the pancreas, which can be classified as acute, acute recurrent, or chronic pancreatitis. However, this condition may be more easily considered based on etiology, as there is a myriad of potential causes of pancreatitis in the pediatric population, from developmental anatomical abnormalities to rising incidence of obesity. Subtypes of pancreatitis include necrotic and hemorrhagic.
2. Acute pancreatitis results from ectopic activation of pancreatic enzymes. In contrast, chronic pancreatitis is a continuing inflammatory process of the pancreas, characterized by irreversible morphologic changes (i.e. calcifications, fibrosis, ductal stricture, dilatation), which may lead to exocrine and endocrine insufficiency.
3. The risk of recurrent pancreatitis in the pediatric population has been estimated at 15–30%. This may lead to chronic pancreatitis, thus highlighting the importance of prompt recognition and treatment of pancreatitis in its acute phase.

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4. Current practice utilizes a staged approach to the treatment of pancreatitis in children, with endoscopic intervention prior to consideration of open surgical techniques.
5. While the need for surgical intervention for pancreatitis in the pediatric population is uncommon, consideration should be made for cases of failed medical and endoscopic management (often with multiple hospitalizations), chronic pain, obstructing stones, and other complications such as pseudocysts and strictures. The goal of surgery in these cases is to improve quality of life, address the pathophysiologic complications of pancreatitis, and to preserve the integrity of the pancreas as an endocrine and exocrine organ.

1. *Background*
    a. Epidemiology
        i. Pancreatitis is an uncommon disease in the pediatric population, and its etiologies are more diverse compared to adults.
        ii. Males and females are equally affected.
        iii. Overall mortality rate of pancreatitis in the general population is 10%, and this increases to 50% with hemorrhagic pancreatitis.
    b. Etiologies
        i. While most cases of acute and chronic pancreatitis are idiopathic, the most common identifiable etiologies of *acute pancreatitis* are listed below:
            1. Abdominal trauma, including non-accidental trauma (NAT, child abuse) and iatrogenic cause following endoscopic retrograde cholangiopancreatography (ERCP)
            2. Biliary disease, including choledocolithiasis, gallstone pancreatitis
            3. Congenital abnormalities
                a. Pancreas divisum
                b. Annular pancreas
                c. Heterotopic pancreas, most commonly in the duodenum
                d. Choledochal cyst
            4. Multisystem disease
            5. Drugs and toxins, i.e., ethanol, L-asparaginase, Valproic acid, Azathioprine, Mercaptopurine, Mesalamine, steroids
            6. Viral infection
        ii. Although there are many shared etiologies between acute and chronic pancreatitis, cases of *chronic pancreatitis* involve a higher representation of genetic conditions, as listed below:

1. Cystic Fibrosis (CFTR mutation, autosomal recessive)
2. Hereditary pancreatitis (PRSS1 mutation: cationic trypsinogen, autosomal dominant)
3. Metabolic disorders, i.e., hypertriglyceridemia
4. Autoimmune pancreatitis
5. Juvenile tropical pancreatitis
6. Chronic fibrosing pancreatitis
7. Pancreatic tumors

2. *Pathophysiology*
   a. Developmental anatomy
      i. Ventral/dorsal pancreatic bud: at 6 weeks *in utero*, the ventral bud rotates posteriorly, to the right, and clockwise, thereby fusing with the dorsal bud.
         1. Ventral bud = uncinate process, inferior portion of head of pancreas
            a. Duct of Wirsung- main pancreatic duct that merges with common bile duct, then drains into duodenum at Ampulla of Vater, via Sphincter of Oddi
         2. Dorsal bud= body, tail, superior portion of head of pancreas
            a. Duct of Santorini- accessory duct that drains into duodenum via accessory papilla
      ii. Annular pancreas
         1. Failure of clockwise rotation of the ventral bud results in duodenal obstruction of the D2 segment by the pancreas or band, which can lead to pancreatitis.
      iii. Pancreas divisum
         1. Failure of fusion of pancreatic Ducts of Wirsung and Santorini results in the majority of pancreatic enzymes draining via the accessory papilla. Stenosis of the Duct of Santorini may also be present, causing pancreatitis.
   b. Endocrine function
      i. Alpha cells secrete glucagon
      ii. Beta cells secrete insulin
      iii. Delta cells secrete somatostatin, vasoactive intestinal peptide (VIP)
      iv. PP or F cells secrete pancreatic polypeptide
      v. Gastrin-producing cells present in fetal pancreas only
   c. Exocrine function: amylase, lipase, trypsinogen, chymotrypsinogen, carboxypeptidase, bicarbonate
      i. Ductal cells secrete HCO3—(contain carbonic anhydrase)
      ii. Acinar cells secrete digestive enzymes (listed above)

    iii. Amylase is the only pancreatic enzyme listed above that is secreted in its active form; the rest require chemical conversion into their active forms.

  d. Hormonal control of enzymatic secretion

    i. Acetylcholine (ACh) and secretin cause an increase in bicarbonate.
    ii. CCK and ACh cause an increase in secretion of pancreatic enzymes.
    iii. Somatostatin and glucagon cause a decrease in pancreatic exocrine function.

  e. In Cystic Fibrosis, the genetically abnormal CFTR membrane protein results in NaCl transport dysfunction, affecting submucosal glands (sinopulmonary disease, Type 1 diabetes) and exocrine epithelial cells. Thick secretions obstruct exocrine ducts, which destroy pancreatic acinar cells. This leads to pancreatitis, and in severe cases, pancreatic insufficiency with malabsorption and failure to thrive.

3. *Clinical presentation*

  a. Classically, pancreatitis presents as acute onset epigastric pain with radiation to the central back. The pain tends to be positional. This may be accompanied by nausea, vomiting, anorexia, abdominal distention, ileus, jaundice (with gallstones or obstruction), fever, tachycardia, and hypotension.
  b. On physical examination, one may find signs of abdominal trauma, tenderness, distention, or peritoneal signs. In cases of hemorrhagic pancreatitis, Grey Turner's sign (bluish discoloration of flanks) and/or Cullen's sign (bluish discoloration of periumbilical region) may be present. This is due to infiltration of blood along subcutaneous planes and the gastrohepatic and falciform ligaments respectively, as the pancreas is a retroperitoneal organ.
  c. Chronic pancreatitis may mimic acute pancreatitis. It typically presents as chronic abdominal pain that is difficult to treat. It may be accompanied by signs of pancreatic exocrine and endocrine insufficiency, such as diabetes with labile blood sugars, steatorrhea, diarrhea, ADEK vitamin deficiency, anorexia, and failure to thrive.

4. *Diagnosis*

  a. Several classification systems for pancreatitis have been utilized in the clinical setting.

    i. The Atlanta classification (revised in 2012) characterizes acute pancreatitis by an early (<2 weeks) and late (>2 weeks) phase, with mild, moderate, and severe cases.
    ii. Specific to pediatrics, Morinville et al. in 2012 described the INSPPIRE criteria for diagnosis of acute pancreatitis with any two of these: abdominal pain, or imaging consistent with pancreatitis, or lipase/amylase 3 times the upper limit of normal.

    iii. Modified Ranson criteria may be used for a pediatric scoring system, though ongoing efforts are being made to explore the utility of current prediction severity tools published in the adult population by adding pediatric systemic inflammatory response syndrome (SIRS) score, age, and weight.

  b. If pancreatitis is suspected based on history and physical examination, amylase and lipase should be measured, and other laboratory values may also be useful in establishing the diagnosis.

    i. Serum amylase and lipase (more specific) tend to be at least 3× the upper limits of normal.
    ii. While amylase is a non-specific indicator of pancreatitis, amylase-P is a laboratory test available at some centers, whose specificity is closer to that of lipase.
    iii. Other lab findings may include: leukocytosis, hyperglycemia, hypocalcemia, hypertriglyceridemia, hypoalbuminemia, hyperbilirubinemia, elevated LFTs, acidosis, glycosuria.
    iv. Sweat chloride and modern genetic testing will be utilized in cases where there is suspicion of syndromic or genetic basis for the illness.

  c. Imaging studies provide evidence of structural changes in the glands or ducts. Transabdominal ultrasonography, CT, MRCP, ERCP, and EUS may be utilized, and provide evidence of acute vs. chronic changes.

    i. Ultrasound (US) is a primary screening tool, and its findings may reveal a diffusely enlarged or edematous pancreas with dilated pancreatic ducts, peri-pancreatic fluid collection, abscess, or pseudocyst.
    ii. CT may be utilized for evaluation of abnormalities found on US; chronic pancreatitis, pancreatic necrosis, complications, pancreatic trauma, or neoplastic disease process. CT findings include an enlarged gland with ill-defined margins, peri-pancreatic fluid collection, areas of decreased or enhanced density, or pseudocyst.
    iii. Magnetic resonance cholangiopancreatography (MRCP) is a non-invasive imaging study of biliary tree and surrounding structures, is more sensitive than CT, and helps delineate an anatomical lesion. In children, this imaging modality frequently requires sedation.
    iv. Endoscopic retrograde cholangiopancreatography (ERCP) is an invasive modality that can help diagnose various pancreatic and biliary anomalies, ductal abnormalities, or obstructions. It can also serve as a therapeutic intervention, i.e., sphincterotomy and stent placement.

5. *Initial management*

  a. The management of acute pancreatitis usually consists of supportive and medical therapy with IV fluid hydration and pain control. Initial management is directed at stabilizing the patient and aggressive hydration measures to help decrease the risk of multi-organ failure. This should include strict measurement of ins and outs, and serial labs as indicated.

b. Bowel rest with parenteral nutrition versus enteral nutrition has been a topic of much debate and research in the adult population. In adults, meta-analyses of practices regarding nutrition in pancreatitis have clearly shown that enteral over parenteral nutrition reduces mortality, incidence of multisystem organ failure, and need for invasive procedures. The data has not been as heavily explored in the pediatric population, but current practice does support the implementation of early enteral feeding, within 24–48 h following admission. In fact, if during the course of a pancreatitis episode the patient develops the desire to eat, this may be a sign that the severe pancreatic inflammation has begun to resolve.

c. In cases of severe pancreatitis, total parenteral nutrition (TPN) may be necessary if the patient cannot tolerate enteral feeds, or in conjunction with feeding.

    i. Jejunal feeds via small bore feeding tube are preferred over TPN due to lower complication rates and lower costs.
    ii. In cases of chronic pancreatitis, low-fat meals and pancreatic enzyme supplements may help restore exocrine enzyme deficiencies.

6. *Treatment*

    a. Pancreatitis prevention

        i. In cases of hereditary pancreatitis or a known predisposition based on genetic markers, it may be useful to supplement diet with fibrates, fish oil, and abstain from alcohol.
        ii. Congenital abnormalities of the pancreas have the potential to cause pancreatitis. However, these conditions are typically diagnosed early in life and their unique interventions will ideally reduce the chances of developing pancreatitis.

            1. Annular pancreas presents with signs and symptoms of duodenal obstruction (nausea, vomiting, abdominal pain; X-ray with double bubble sign). Options for surgical intervention are duodenojejunostomy or duodenduodenostomy *without* resection of the pancreas.
            2. Pancreas divisum may present with pancreatitis, but most cases are asymptomatic. Both diagnosis and treatment utilize ERCP with sphincteroplasty. In this procedure, canalization of the minor papilla will reveal a long, large duct of Santorini, while canalization of the major papilla will reveal a short duct of Wirsung.

    b. Medical treatment

        i. Diabetes screening and management, including yearly glucose and hemoglobin A1C
        ii. Early identification and treatment of fat-soluble vitamin deficiencies

            1. Patients typically require antacid medication in addition to pancreatic enzyme supplementation to assist with breakdown.

- c. Pancreatitis procedures
    - i. ERCP or EUS may be useful in mitigating threats of continued pancreatic damage from obstruction at the common bile duct or pancreatic duct.
        1. For example, ERCP is the mainstay of treatment for gallstone pancreatitis with obstruction stones, as evidenced by persistently elevated bilirubin/LFTs, and dilated ducts on US or MRCP.
        2. Patients with gallstone pancreatitis require interval cholecystectomy when pancreas inflammation has resolved.
    - ii. Surgical management of pancreatitis is rarely required, and it is reserved for cases of chronic, relapsing pancreatitis, failed conservative medical therapy, intractable pain, impaired nutrition, and narcotic addiction.
    - iii. Surgical reconstructive options include distal pancreatectomy with Roux-en-Y pancreaticojejunostomy (Duval procedure), and lateral pancreaticojejunostomy (Puestow procedure) with or without anterior resection of the pancreatic head (Frey procedure).
    - iv. Laparoscopic or open surgical debridement is indicated for infected pancreatic abscess or necrosis, and emphysematous pancreatitis.
    - v. Video-assisted retroperitoneal debridement (VARD procedure) has been developed more recently in the treatment of chronic severe pancreatitis in adults, as a blend of endoscopic retroperitoneal necrosectomy and open surgery.
    - vi. In patients with Cystic Fibrosis, pancreas transplantation may be indicated, in addition to double lung transplantation.

7. *Outcomes*
    - a. Early complications—days to weeks
        - i. Development of ARDS due to pro-inflammatory mediators, including phospholipase, causing capillary leakage. This non-cardiogenic pulmonary edema can usually be diagnosed with chest X-ray (CXR) and may require intubation for ventilator support.
        - ii. Pancreatic saponification due to enzymatic breakdown of the tissue can lead to pancreatic necrosis. The saponification reaction also results in hypocalcemia. Diagnose with ionized calcium level, and replete with IV calcium.
        - iii. Infected pancreatic necrosis with systemic inflammatory response syndrome (SIRS)
            1. This is diagnosed by CT-guided tissue biopsy (performed to rule out malignancy) and culture (to guide the use of antibiotics) and ultimately requires surgical debridement as described in treatment.
            2. Carbapenems (Imipenem, Meropenem) are preferred over ciprofloxacin in the treatment of infective pancreatitis.

    iv. Ascites and pleural effusions are due to retroperitoneal leakage of pancreatic fluid. These collections seen on US and CXR should not be drained unless there is a concern for infection, as they will resolve as the disease process is addressed.

b. Late complications—weeks to months
    i. Pancreatic pseudocyst formation
        1. A pseudocyst is *not* epithelial-lined; commonly forms in the head of the pancreas
        2. Can cause mass effect resulting in early satiety, abdominal fullness, and even small bowel obstruction
        3. Typically resolve spontaneously
        4. Treatment is based on size and timing:
            a. If the pseudocyst is <6 cm AND has been present for >6 weeks, watch and wait.
            b. If it is >6 cm OR >6 weeks, perform drainage with biopsy (EGD or percutaneous).
            c. Once the pseudocyst has formed a mature wall, OR if growing over time, perform percutaneous vs. open cystogastrostomy.
    ii. Pancreatic fistulas usually arise from complications of pancreatic operations.
        1. Spontaneous closure in low output fistulas (<200 cc/day)
        2. Treatment involves NPO with TPN and octreotide, but ERCP with pancreatic stenting may be needed if fistula fails to close.
    iii. Pancreatic abscesses are treated with surgical drainage and antibiotics.
    iv. Mechanical obstruction of duodenum and common bile duct
    v. Splenic vein thrombosis with portal hypertension
        1. Gastric varices may form as collaterals. Splenectomy is indicated for isolated bleeding varices.
    vi. Splenic artery pseudoaneurysm
    vii. Pancreatic insufficiency
        1. This is an unfortunate sequela of chronic pancreatitis, and typically exocrine function of the pancreas is more impacted than endocrine function.
        2. Adherence to a diet high in carbohydrates and protein, and low in fat along with enzymatic supplementation is important, but ultimately, these patients need pancreas transplantation.

## Additional Notes

The rising incidence of pancreatitis in children is thought to be due to increasing obesity and drug abuse rates in this population, and also perhaps due to improved detection strategies. While classic causes of pancreatitis in adults like alcohol, gallstones, iatrogenic trauma, and medication reactions may be seen in pediatric cases, the consideration of congenital etiologies- from anatomic anomalies to genetic disorders of metabolism- significantly expands the differential. Although over time several classification models have been developed for the diagnosis of this condition, a laboratory value of lipase 3× the upper limit of normal remains a strong indicator of pancreatitis. Imaging by CT is not necessary if symptoms and lab values correspond to a diagnosis of pancreatitis, as early pancreatitis may not be visualized on such imaging; rather, obtain confirmatory imaging to assist with diagnosis in cases where lab values are equivocal. Although the pancreas is poorly visualized on ultrasound, this test may be useful in cases of gallstone pancreatitis for visualization of the gallbladder. Similarly, MRCP is useful to identify certain etiologies of pancreatitis, including stricture and malignancy.

Necrotizing pancreatitis has historically resulted in high complication rates and mortality, which has brought about the development of what is known as the "step up pathway," or staged approach. Even within this model, there is an ongoing debate around the myriad options of endoscopic, laparoscopic, open, and blended techniques that are being utilized to address such severe cases of pancreatitis. When medical treatment fails, often therapeutic endoscopy is first-line interventional treatment and is intended to serve as source control and mitigation of sepsis. Options include endoscopic cystgastrostomy with or without stent or pigtail drain placement. Depending on etiology of pancreatitis, presence of anatomical abnormalities, and gene mutation status, appropriate surgical intervention may be explored next. If endoscopic treatment is unsuccessful in mitigating disease, definitive procedures in the step-up approach include pseudocyst drainage, more specifically cystogastrostomy or cyst-enterostomy requiring Roux-en-Y reconstruction; partial pancreatectomy, total pancreatectomy with islet autotransplantation; and longitudinal pancreaticojejunostomy (Puestow procedure) with or without anterior resection of the pancreatic head (Frey procedure). The VARD procedure (video-assisted retroperitoneal debridement) was described in 2007 by Van Santvoort et al. as a blend of endoscopic retroperitoneal necrosectomy and open surgery in the step-up approach to necrotizing pancreatitis in adults, using a left subcostal incision to remove necrotic pancreatic tissue and laparoscopic identification of deeper areas of necrosis, followed by continuous lavage of the retroperitoneum in the post-operative period. Ultimately, the step-up approach aims to achieve long-term pain relief and preserve maximal pancreatic function while minimizing surgical trauma in critically ill patients. Fortunately, this severity of disease progression is seen much more rarely in the pediatric population.

During the novel coronavirus disease 19 (COVID-19) pandemic, some attention has been drawn to understand the extrapulmonary manifestations of this disease, including that of acute pancreatitis. While a causal relationship between pancreatitis and COVID-19 has not been proven, temporal association between the two is identified in several case reports in adults, and a few in children to date. In these pediatric case reports, adolescent patients developed symptoms of acute pancreatitis within 1 week of suffering from COVID-19 illness. As development of pancreatitis has been observed in a constellation of viral diseases (including measles, mumps, EBV, Coxsackievirus), a connection to immune-mediated pancreatic injury or angiotensin-converting enzyme 2 involvement in the setting of COVID-19 infection may be plausible, though this idea requires further investigation.

## Study Questions

1. A 6-year-old male with past medical history significant for prematurity and NEC is being evaluated in the PICU for newly diagnosed gallstone pancreatitis. He is severely dehydrated and requires aggressive pain control measures. What is the best approach in regards to nutritional management of the patient?

    a. Enteral feeds in 1 week
    b. Enteral feeds in 24–48 h
    c. TPN immediately
    d. Enteral feeds immediately

    *Answer: (b.) Enteral feeds in 24–48 h*. Historical practice in the medical management of acute pancreatitis stressed the importance of bowel rest in order to rest the exocrine pancreas to prevent further inflammation and local destruction. More recently, practice has shifted toward early initiation of enteral feeding with positive outcomes in these patients.

2. On hospital day #10 in a 16-year-old male admitted with abdominal pain secondary to hereditary pancreatitis, the patient develops fever and tachycardia with leukocytosis. Interval CT scan demonstrates development of gas pockets within the body of an inflamed pancreas with areas of necrosis. What is the most appropriate next step in the treatment of this patient?

    a. Immediate percutaneous drainage
    b. VARD procedure
    c. Transfer to PICU with close monitoring including serial abdominal exams
    d. Initiate IV antibiotics alone

    *Answer: (a.) Immediate percutaneous drainage*. Although initiation of antibiotics is warranted, given the patient's fever and elevated WBC count, it is also necessary to achieve source control in this case of infected pancreatic necrosis. At the time of drain placement, tissue biopsy and culture are useful to rule out malignancy and guide antibiotic selection.

3. A 5-month-old male presents to the emergency department with fussiness, abdominal distention, non-bilious vomiting, and anorexia that began 1 day ago. Ultrasound demonstrates a proximal narrowing past the pylorus. What laboratory studies may help distinguish the likely diagnosis from other conditions that may present with similar symptoms?

    a. Electrolytes
    b. Blood smear
    c. Amylase, lipase
    d. Liver function tests

    *Answer: (c.) Amylase, lipase*. The differential in an infant with these symptoms on presentation is wide, but description of the vomitus and imaging are helpful in working toward an appropriate diagnosis. Abnormally elevated amylase or lipase 3× above the upper limit of normal, along with the above US findings, suggests a congenital abnormality resulting in pancreatitis in this patient. Electrolyte abnormalities may be seen in hypertrophic pyloric stenosis, along with projectile bilious vomiting. Blood smear may be revealing in a patient with a blood-borne illness, whether viral or hereditary. Abnormal LFTs are typically accompanied by jaundice in cases of biliary disease.

## Further Reading

Diesen DL. Acute pancreatitis, Chronic pancreatitis, Pancreatitis procedures. Pediatric Surgery NaT, American Pediatric Surgical Association. 2020. APSA Webapp. www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT

Fialkowski E, Pryor HI, Fischer A, et al. Congenital duodenal obstruction. Pediatric Surgery NaT, American Pediatric Surgical Association. 2020. APSA Webapp. www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829038/all/Congenital_Duodenal_Obstruction

Gariepy CE, Heyman MB, Lowe ME, et al. The causal evaluation of acute recurrent and chronic pancreatitis in children: consensus from the INSPPIRE Group. J Pediatr Gastroenterol Nutr. 2017;64(1):95–103.

Morinville VD, Husain SZ, Bai H, et al. Definitions of pediatric pancreatitis and survey of present clinical practices. J Pediatr Gastroenterol Nutr. 2012;55(3):261–5.

Sacco Casamassima MG, Goldstein SD, Yang J, et al. The impact of surgical strategies on outcomes for pediatric chronic pancreatitis. Pediatr Surg Int. 2017;33(1):75–83.

Samies NL, Yarbrough A, Boppana S. Pancreatitis in pediatric patients with COVID-19. J Pediatr Infect Dis Soc. 2021;10(1):57–9.

Stylianos S, Cofer BR, Bass KD. Pancreatic trauma. Pediatric Surgery NaT, American Pediatric Surgical Association. 2020. APSA Webapp. www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829089/all/Pancreatic_Trauma

Suzuki M, Saito N, Naritaka N, et al. Scoring system for the prediction of severe acute pancreatitis in children. Pediatr Int. 2015;57(1):113–8.

Van Santvoort HC, Besselink MG, Bakker OJ, et al. A step-up approach or open necrosectomy for necrotizing pancreatitis. N Engl J Med. 2010;362(16):1491–502.

Way WW, Levin MJ, Abzug MJ, et al. Current diagnosis and treatment: pediatrics. 25th ed. New York: McGraw Hill; 2020.

# Gallbladder Disease

**Samir Narula, Pratibha Vemulapalli, and Brian Gilchrist**

**Key Points**

1. Conditions that predispose to gallbladder pathology are obesity, hemolytic disease, sickle cell disease, spherocytosis, parenteral nutrition, ileal resection, Crohn's disease, shock/sepsis, Type 3 progressive familiar intrahepatic cholestasis, and defect in ABCB4 gene.
2. Laparoscopic cholecystectomy is indicated for cholecystitis, symptomatic gallstones, gallstone pancreatitis, and biliary dyskinesia.
3. Precise knowledge of biliary tract anatomy and variations is necessary to avoid intraoperative complications.

Surgery of the biliary tract demands an intrepid surgeon; the intrepid surgeon must have a thorough understanding of its anatomy and myriad variants. Grave damage to a patient can be done by those who are not technically adept and knowledgeable regarding biliary surgery. Pediatric cholecystectomy is an operation that is being done with greater frequency today for reasons that are many and varied. However, the obesity of our pediatric population is the major causal factor in this increased need for gallbladder surgery among children.

Another major reason for this increase in cholecystectomy is detection of biliary dyskinesia through the use of HIDA scan. Children have gallstones less frequently

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than adults, but when they do, pigment stones, rather than cholesterol stones, are more common. This too is changing due to diet indiscretion in the adolescent population.

1. Epidemiology
   A. Pediatric biliary disease in increasing in incidence. Approximately 2% of children have gallstones, and a greater number have biliary disease without stones. After puberty, females have quadruple the incidence of gallstones compared to males.
2. Anatomy (Fig. 1)
   A. The gallbladder lies in the gallbladder fossa within the ventral surface of the liver. The gallbladder is divided into the fundus, body, and neck (referred to as the infundibulum).
   B. The cystic duct joins the common hepatic duct to form the common bile duct.
   C. Blood supply:
      i. The cystic artery arises from the right hepatic artery. Cystic arteries are quite variable and may arise from the right, left or common hepatic arteries.

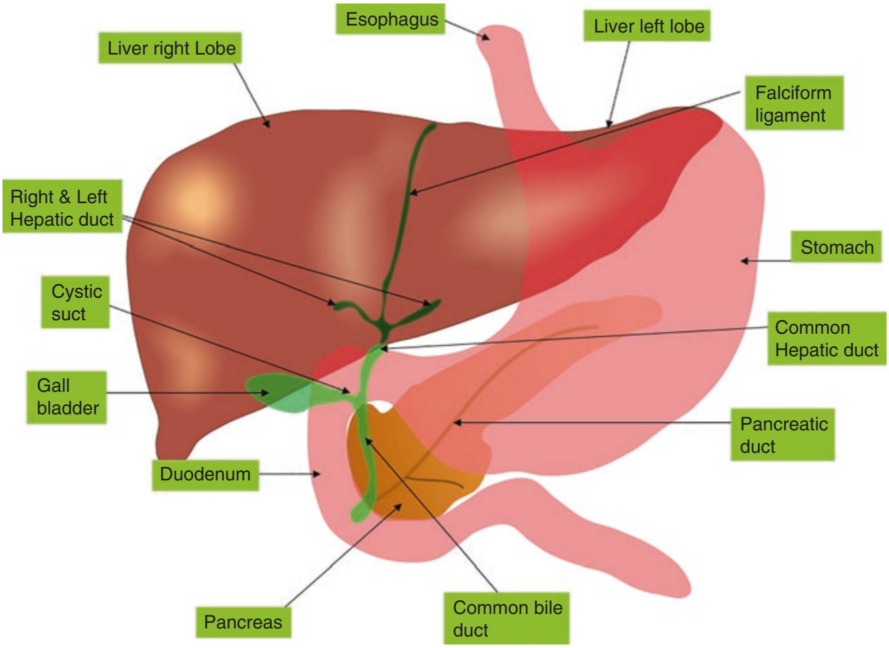
Esophagus
Liver left lobe
Liver right Lobe
Falciform ligament
Right & Left Hepatic duct
Stomach
Cystic suct
Common Hepatic duct
Gall bladder
Pancreatic duct
Duodenum
Pancreas
Common bile duct

**Fig. 1** Gallbladder anatomy. (Source: Jiju Kurian Punnoose, https://commons.wikimedia.org/wiki/File:Anatomy_of_liver_and_gall_bladder.png , Accessed 30 Nov 2021, used via Creative Common Share Alike 4.0 International license: https://creativecommons.org/licenses/by-sa/4.0/)

D. The critical anatomy in gallbladder surgery is defining the boundaries of the triangle of Calot:

  i. The common hepatic duct on the left.
  ii. The cystic duct on the right.
  iii. The liver superiorly.

3. Pathophysiology

  A. The gallbladder functions to store bile during periods of fasting. The gallbladder concentrates bile. During consumption of a meal particularly due to fat, the duodenum secretes cholecystokinin, which causes the gallbladder to contract and the sphincter of Oddi (the sphincter between the common bile duct and duodenum) to relax.
  B. Cholelithiasis: The presence of gallstones can be asymptomatic or associated with abdominal pain. Bile is usually liquid, but unbalanced proportions of cholesterol, bile salts, and lecithin lead to precipitation of gallstones. Stress, shock, and dehydration can lead to concentration of bile and gallstones.

    a. Types of gallstones: The color reflects content, and usually there will be one type of stone present. Cholesterol stones are most common in adults, but children have greater incidence of pigment stones.

      a. Black (48%): pigment stones (calcium bilirubinate) present in hemolysis and parenteral nutrition.
      b. Yellow (21%): from supersaturation of bile with cholesterol.
      c. Protein stones (5%).
      d. Calcium carbonate (4%).
      e. Brown (3%): associated with cholangitis and bile stasis, made of bilirubin, fatty acids, and calcium.

  C. Cholecystitis (inflammation of the gallbladder): occurs when gallstone (or other process) obstructs the cystic duct, creating pressure, stasis, distention, vascular congestion, edema, and ischemia of gallbladder.
  D. Gallstone pancreatitis: occurs when stones from gallbladder pass through common bile duct and obstruct pancreatic duct.
  E. Biliary dyskinesia: Occurs when there is impaired or painful emptying of the gallbladder after a meal without the presence of gallstones. The etiology of this process is not known, but may be due to a long cystic duct, tortuous valves in duct, low junction of the cystic duct and common bile duct, or a visceral hypersensitivity of the gallbladder.
  F. Biliary hyperkinesis: Defined as an ejection fraction of >70% or recreation of symptoms with injection of cholecystokinin.
  G. Associated conditions:

    a. Increased incidence of cholelithiasis is seen in hemolytic disease, obesity, use of parenteral nutrition, after ileal resection, Crohn's disease, shock/sepsis, and pregnancy.

   b. Patients with sickle cell disease have double the incidence of gallstones. Half of children with sickle cell disease have gallstones by the time they are adults. The gallstones seen in sickle cell disease are pigmented stones.
      (a) Spherocytosis with increased turnover of red blood cells leads to pigment stones.
      (b) Inherited conditions predisposing to cholelithiasis (cholesterol stones) are progressive familiar intrahepatic cholestasis, Type 3, and defect in ABCB4 gene.

4. Clinical presentation
   A. Biliary colic: A sharp stabbing pain in the right upper quadrant that comes and goes. Pain may be related to dietary intake. It can be associated with nausea and symptoms of reflux. Biliary colic can accompany gallstones and biliary dyskinesia. An attack of pain lasting greater than 6 h is suggestive of cholecystitis.
   B. Patients may also have avoidance of fatty foods, anorexia, and weight loss as they learn that certain foods worsen their symptoms.
   C. Many patients with cholesterol stones are obese.
   D. Charcot's triad: jaundice, fever, and right upper quadrant pain. Suggests choledocholithiasis.
   E. Reynold's pentad: components of Charcot's triad plus shock and altered mental status. Suggests ascending cholangitis.
   F. Murphy's sign: This physical exam finding is elicited by palpating the liver edge under the right costal margin. The patient is asked to inhale deeply, and if this causes right upper quadrant pain, Murphy's sign is present. A sonographic Murphy's sign is when the sonographer compresses the gallbladder with the ultrasound probe and elicits pain.

5. Diagnosis
   A. Abdominal sonography (specifically the right upper quadrant): can detect gallstones and choledocholithiasis. Cholecystitis is identified by the presence of pericholecystic fluid and thickening of the gallbladder wall. Ultrasound also can detect enlargement of the common bile duct or the intrahepatic bile ducts, which indicates distal obstruction. Sonography is best when patients are NPO because there is less bowel gas present to interfere with ultrasound transmission. Sonography can also detect alternate pathology.
   B. Lab tests: patients presenting with right upper quadrant pain should undergo complete blood count to detect leukocytosis, electrolytes to detect dehydration and electrolyte abnormalities, liver function tests, gamma-glutamyl transferase to detect hyperbilirubinemia, hepatitis panel (A–B–C), and elevated alkaline phosphatase. Testing amylase and lipase will detect pancreatitis, and a urinalysis will differentiate a urinary tract infection or urinary stone from biliary disease.

   C. Abdominal X-ray is useful to discern bowel gas pattern when patients are vomiting, and rarely gallstones with high calcium content will be visible.
   D. Magnetic resonance cholangiopancreatography (MRCP) can identify biliary ductal obstruction when lab tests show hyperbilirubinemia or pancreatitis. Younger children will require sedation or an anesthetic to accomplish MRCP.
   E. Hepatobiliary iminodiacetic acid (HIDA) scan with gallbladder ejection fraction (GBEF): HIDA scan will demonstrate ability of the liver to conjugate bile, patency of the cystic duct, and patency of the common bile duct. Additionally, a gallbladder ejection fraction (percentage of tracer in gallbladder excreted 1 h after gallbladder is stimulated with cholecystokinin analog) can be measured. Values less than 35% are considered diagnostic of biliary dyskinesia. Values >70% associated with pain may indicate a hyperkinetic gallbladder, and even if the GBEF is normal, if the patient has pain with cholecystokinin analog administration that mimics pain they feel with meals, they may benefit from cholecystectomy.
   F. Computed tomography: Not typically used to diagnose biliary disease and carries the risk of ionizing radiation. However, gallstones can be visible on CT scan.

6. Initial stabilization

   A. Patients with cholangitis and or pancreatitis are in danger of sepsis and shock so need immediate fluid resuscitation and antibiotics. Some may require intensive care unit monitoring.
   B. Patients with cholecystitis need fluid and antibiotics.
   C. Pain control is given for the biliary colic.
   D. Patients who have evidence of biliary ductal obstruction or gallstone pancreatitis need to have the common bile duct cleared. For many patients, the choledocholithiasis will clear spontaneously; this will be evident in improving symptoms and decreasing serum bilirubin. MRCP is useful to define the presence of common bile duct stones. For those with common bile duct stones, ERCP should be performed prior to cholecystectomy, followed by cholecystectomy within 24 h.
   E. Patients with intermittent pain, who are afebrile, and can tolerate oral hydration can be scheduled for elective cholecystectomy as an outpatient.

7. Treatment

   A. Patients with biliary colic can be considered for a trial of medication for alternate possible diagnoses, such as histamine 2 blockers, proton pump inhibitors, laxatives, and antispasmodics, but if the cause of the pain is biliary, these are unlikely to help.
   B. Asymptomatic gallstones in children can be observed but should be checked for resolution with sonography in 6 months. Ursodeoxycholic acid can be administered. If there is a known underlying condition responsible for the gallstones, such as hemolytic disease, they are unlikely to resolve. Gallstones in children younger than eight without a known etiology are more likely to

resolve. If at any point the gallstones become symptomatic, or if sequelae such as cholecystitis or obstruction occur, observation should be abandoned.

C. Laparoscopic cholecystectomy is indicated for cholecystitis, symptomatic gallstones, gallstone pancreatitis, and biliary dyskinesia. If there is any question of choledocholithiasis at the time of cholecystectomy, an intraoperative cholangiogram can be performed. Patients can undergo cholecystectomy as an outpatient. Patients with sickle cell anemia should be admitted the day before surgery for hydration and transfusion when needed. After surgery they should be well hydrated to prevent a sickling crisis from the stress and fluid shifts of operation.
D. Laparoscopic cholecystectomy can be converted to open cholecystectomy if needed for safe completion of the operation.

8. Outcomes

A. Possible complications of cholecystectomy: common bile duct injury, hemorrhage, bile leak, biloma, diarrhea, gas.
B. Common bile duct strictures can be treated with endoscopic balloon dilation.
C. Injury or inadvertent clipping of the common bile duct during cholecystectomy if recognized can be treated by direct repair with T-tube placement. Late recognized injuries are best treated by hepaticojejunostomy.
D. Nearly all patients with symptomatic gallstones have resolution of symptoms, while 60–80% of patients with acalculous biliary colic and biliary dyskinesia are improved with cholecystectomy.

## Additional Notes

Although pediatric surgeons are well trained in biliary surgery, traumatic injury to the biliary tract during gallbladder surgery necessitates careful identification of the biliary anatomy in the triangle of Calot. One must be prepared to perform cholangiography, open surgery, or subtotal cholecystectomy to avoid injuries. Poor outcomes from inadequate surgery in this setting can have long-term grave morbidities.

## Study Questions

1) Question 1: True or False: HIDA scan is the standard for evaluation of biliary dyskinesia?

   a. True
   b. False

Answer: (a) True. The HIDA scan is a dynamic study to evaluate for biliary dyskinesia. An ejection fraction <35% is considered diagnostic of biliary dyskinesia.

2) True or False: The arterial anatomy of the biliary tract is anomalous in 40% of cases?

   a. True
   b. False

Answer: (a) True. Anomalous biliary anatomy is common. Two common anomalies with dire consequences if unrecognized are a displaced right hepatic artery which can be mistaken for the cystic artery and the absence of a cystic duct.

## Further Reading

Brazelli M, Cruikshank M, Kilonzo M, Ahmed I, et al. Clinical effectiveness and cost-effectiveness of cholecystectomy compared with observation/conservative management for preventing recurrent symptoms and complications in adults presenting with uncomplicated symptomatic gallstones or cholecystitis: a systematic review and economic evaluation. Health Technol Assess. 2014;18(55):1–101.

Gazzetta J, Fan B, Bonner P, Galante J. Cholecystectomy for biliary hyperkinesia. Am Surg. 2019;85(2):219–22.

Huckaby L, Timmapuri S, Prasad R. Biliary hyperkinesia: a potentially surgically correctable disorder in adolescents. J Pediatr Surg Case Rep. 2013;1(9):314–6.

Kapoor VK. Bile duct injury: when? what? who? J Hepatobiliary Pancreat Surg. 2007;14(5):476–9.

Santucci NR, Hyman PE, Harmon CM, Schiavo JH, et al. Biliary dyskinesia in children: a systematic review. J Pediatr Gastroenterol Nutr. 2017;64(2):186–93.

Svensson J, Makin E. Gallstone disease in children. Semin Pediatr Surg. 2012;21(3):255–65.

Wesdorp I, Bosman D, de Graaff A, Aronson D, et al. Clinical presentations and predisposing factors of cholelithiasis and sludge in children. J Pediatr Gastroenterol Nutr. 2000;31(4):411–7.

# Acute Scrotum and Testicular Torsion

**Jasmine I. Kashkoush and Robert Steckler**

**Key Points**

1) Always consider testicular torsion during acute scrotum presentation. Imaging should never delay your management if your clinical suspicion of testicular torsion is high.
2) The viability of the testicle is related to the degree of spermatic cord torsion and duration of torsion, with most testicles salvageable if duration <6–8 h.
3) Following testicular trauma, a contusion or hematocele can be treated conservatively but testicular rupture requires surgical repair.

Acute scrotum refers to sudden onset scrotal pain and/or swelling. This must be evaluated immediately to rule out a surgical emergency [2, 3].

1. Differential diagnoses (Fig. 1)
    a. Testicular torsion: All acute scrotum presentations should be considered testicular torsion until proven otherwise.
    b. Appendix testis/epididymis torsion
    c. Epididymitis/orchitis
    d. Inguinal hernia/hydrocele

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- Torsion of testicular or paratesticular appendage: Appendix testis or appendage of epididymis, paradidymus, or vas abberans
- Testicular torsion: Intravaginal (acute or intermittent) or extravaginal
- Epididymitis: Infectious (urinary tract infection, sexually transmitted disease, viral), sterile, or traumatic
- Scrotal edema or erythema: Diaper dermatitis, insect bite, skin lesions, idiopathic scrotal edema
- Orchitis: Associated with epididymitis (with or without abscess), Vasculitis such as Henoch-Schönleinpurpura, or viral (mumps)
- Trauma: Hematocele, scrotal contusion,or testis rupture.
- Hernia or hydrocele: Inguinal hernia, hydrocele, associated with abdominal pathology (appendicitis, peritonitis, etc.)
- Varicocele
- Mass: Cystic dysplasia, tumor of testis, Epididymal cyst, spermatocele, epididymal tumor, other paratesticular tumor
- Musculoskeletal pain from inguinal tendonitis or muscle strain
- Referred pain: Ureteral calculus, ureteral anomaly

**Fig. 1** Differential diagnosis of acute scrotal pain (Adapted from: Partin et al. 2020 [1])

   e. Varicocele
   f. Traumatic contusion, hematocele, rupture
   g. Testicular/scrotal mass
   h. Henoch–Schoenlein purpura
   i. Referred pain (e.g., ureteral obstruction)
   j. Idiopathic scrotal edema, insect bite, vasculitis
   k. Cellulitis, Fournier's gangrene

2. Clinical presentation
   a. Age
      i. Infancy to puberty: appendage torsion most common
      ii. Perinatal and pubertal periods: epididymitis and testicular torsion more common
   b. Onset, quality, duration—sudden versus indolent
   c. Precipitating/relieving factors: relation to activity/trauma
   d. Prior similar episodes
   e. Associated symptoms such as fever, nausea/vomiting, hematuria, dysuria, discharge
   f. Recent illnesses
   g. Medical and surgical history
   h. Sexual history

3. Physical examination
   a. Symptoms may be referred to the scrotum therefore evaluation should include complete abdominal, including back/flank in addition to the focused genitourinary examination.

b. Penile examination for circumcision status or other meatal abnormalities which may predispose to urinary infections.
c. Scrotal exam
    i. Skin
        1. Examine for erythema, bruising, warmth, tenderness, swelling, crepitus, discharge.
        2. Local erythema, warmth, crepitus, or discharge raises concern for a scrotal skin/subcutaneous infection.
        3. Cremasteric reflex: Normal reflexive contraction of the hemiscrotum with light stroke of the ipsilateral inner thigh in patients >2yo—absence of this reflex raises concern for testicular torsion.
        4. Blue dot sign: Torsion of testicular appendage may produce blue discoloration seen through scrotal skin stretched over upper pole of the affected testicle.
    ii. Testicles
        1. Examine normal and affected sides for tenderness, firmness, size, position, lie, and masses.
            a. Localize site of maximal tenderness.
            b. Firm testicle, horizontal lie, and elevated position in scrotum are all concerning signs for testicular torsion.
            c. Loss of testicular contour with history of trauma may indicate rupture of the tunica albuginea.
            d. Testicular masses rarely cause pain, but this may represent initial symptom.
    iii. Spermatic cord
        1. Check for palpable mass along the inguinal canal which may indicate inguinal hernia; check for reducibility.
        2. If “bag of worms” appreciated within spermatic cord, evaluate patient in standing and supine positions for presence and resolution of varicocele.

4. Diagnosis
    a. Imaging
        i. If high clinical suspicion for testicular torsion, imaging is unnecessary, and will only serve to prolong warm ischemia time and delay definitive treatment.
        ii. Scrotal ultrasonography (US) is easily performed and reveals critical information about testicular size, homogeneity, blood flow, presence of fluid collections, and masses.

            1. Lack of blood flow within testicular parenchyma or weak flow only seen peripherally concerning for testicular torsion.
                a. Sensitivity of color Doppler US for low/absent blood flow in torsion cases only 63–90% in studies
                b. May be difficult to perform and interpret in small children
            2. Heterogeneous parenchyma throughout the testicle also concerning for necrosis associated with testicular torsion
            3. Enlarged testicular appendage without flow, adjacent to a hyperemic epididymal head can be seen in cases of appendix torsion if looked for but is commonly misdiagnosed as epididymitis.
            4. Enlarged or hyperemic epididymis suggestive of epididymitis
            5. Scrotal/testicular abscess may appear as heterogeneous fluid collection with or without septations.
            6. Hematocele or testicular rupture can be seen following trauma.
        iii. KUB or CT may reveal air in the scrotum if concerned for Fournier's gangrene.
        iv. Consider renal US if history and physical suggestive of ureteral obstruction
    b. Laboratory data
        i. Urinalysis to examine for presence of bacteria, WBCs, leukocyte esterase, and/or nitrites as possible indicators of a urinary infection
        ii. Consider STD testing in sexually active male.
        iii. Tumor markers (bHCG, AFP, and LDH) should be ordered in the patient with a testicular mass.
    c. Further information regarding the evaluation and management of inguinal hernias, hydroceles, and varicoceles can be found in their respective chapters.

5. Treatment
    a. If testicular torsion is suspected or cannot be ruled out, patient should undergo emergent scrotal exploration—though US is routinely performed prior to evaluation by the urologic surgeon.
        i. Torsed testicle should be detorsed and allowed several minutes for flow to return to determine viability; may use intraoperative Doppler to determine viability if color does not completely return to the testicle (Fig. 2).
            1. Viability related to degree of spermatic cord torsion and duration of torsion, with most testicles salvageable if duration <6–8 hr
            2. If testicle viable, should be fixed in three points to dartos fascia
            3. If non-viable, simple scrotal orchiectomy should be performed.
        ii. While waiting to assess affected side, contralateral testicle should be exposed, examined, and fixed to prevent future torsion

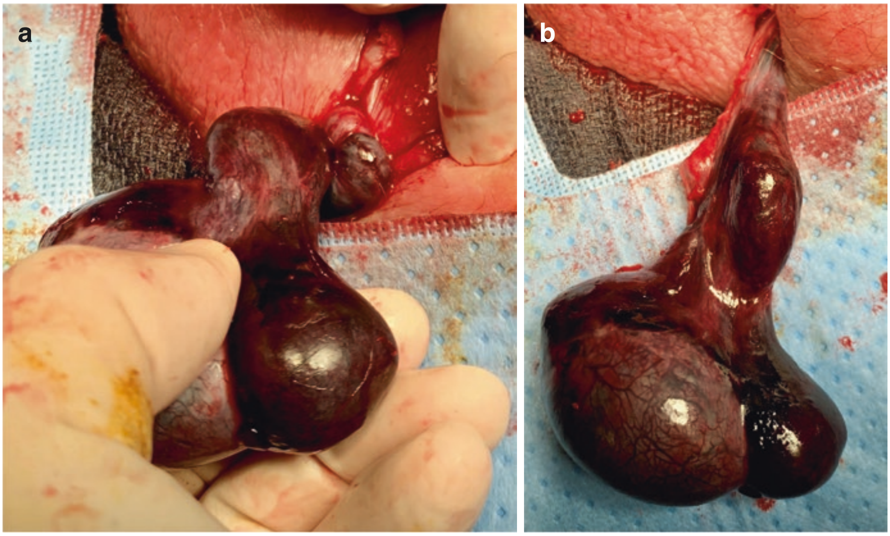

**Fig. 2** Testicular torsion at surgical exploration. (**a**) Testicle with torsion of spermatic cord. (**b**) Testicle after untwisting torsion of spermatic cord. (Source: Dr. Robert Steckler)

b. Alternatively for testicular torsion, bedside manual detorsion can be attempted though sedation may be required—this is a temporizing measure and in NOT a substitute for surgical exploration.
    i. During manual detorsion of the testes, each testicle should be untwisted as if "opening a book"—for the right testicle, untwist the torsion in a counter-clockwise direction [2]. For the left testicle, untwist the torsion in a clockwise direction
    ii. Series have demonstrated that testicular torsion may occur in non-traditional rotational direction in up to one-third of cases.

c. In perinatal cases of testicular torsion, the rotation of the spermatic cord is typically extravaginal and present for an extended period of time (likely occurred before birth). Management is controversial.
    i. Very low chance of testicular salvage therefore some defer surgery until patient is older and able to tolerate anesthetic for scrotal exploration—if surgery is delayed, must acknowledge the risk of metachronous perinatal torsion.
    ii. In instances when an initial scrotal examination is documented as normal and an acute change is noted, and testicular torsion is suspected, emergent surgery is indicated.

d. With normal US findings following an acute presentation but history is suggestive of testicular torsion/detorsion episodes, should consider prophylactic bilateral scrotal orchidopexies for intermittent testicular torsion.

- e. Appendix testis torsion is best treated conservatively with rest and anti-inflammatories.—commonly misdiagnosed as epididymitis by US
- f. Cases of epididymitis/orchitis without abscess formation may be treated conservatively
    - i. Antibiotics should only be used if clinical history or lab evaluation concerning urinary infection (e.g., history of UTIs, suspicious UA)
        1. Start with empiric antibiotics and adjust per urine culture sensitivities—based on patient's age and status of sexual activity.
        2. In children not yet sexually active, consider dysfunctional voiding as the source for the ascending infection.
    - ii. Empiric treatment for chlamydia/gonorrhea should be considered if the patient is sexually active.
    - iii. Non-bacterial epididymitis (idiopathic or viral) treated conservatively with rest and anti-inflammatories.
- g. If concern for scrotal skin/subcutaneous infection, treatment should be directed at the severity and presence/absence of abscess.
    - i. Cellulitis alone should be treated with antibiotics.
    - ii. Scrotal wall abscess may be drained at bedside or in the OR based on patient tolerability and abscess complexity.
- h. With a diagnosis of incarcerated or strangulated inguinal hernia, the patient should undergo immediate surgical exploration (see inguinal hernia chapter).
- i. Following testicular trauma, contusion or hematocele can be treated conservatively but testicular rupture requires surgical repair.
    - i. On scrotal exploration, rupture appears as disrupted tunica albuginea with varying degrees of hemorrhage and extruded seminiferous tubules.
        1. If salvageable, extruded tubules should be debrided and tunica closed.
        2. If not salvageable, simple orchiectomy should be performed.
- j. Patients with a testicular mass should undergo semi-urgent inguinal exploration with partial vs radical orchiectomy as indicated by pathology findings.

6. Follow-up
    - a. After any surgical management of the acute scrotum, the affected testis (if salvaged) and contralateral testicle should be followed longitudinally to assess for atrophy.
        - i. May offer testicular prosthesis after orchiectomy if patient interested
    - b. Patients treated for epididymitis/orchitis should be followed closely for resolution of infection.
        - i. Consider abscess formation if patient has persistent infectious or scrotal symptoms.

   - ii. If suspected viral orchitis does not resolve with conservative measures, consider infectious disease consult as atrophy can result if untreated.
   - iii. Repeat scrotal US may be considered to rule out an underlying testicular mass.
   - iv. Further evaluation for genitourinary abnormalities which may predispose to urinary infection as clinically indicated and in recurrent cases
     1. Renal US, VCUG, cystoscopy as indicated

- c. After treatment of scrotal skin/subcutaneous infection, patients' scrotal exam should be followed closely for improvement with further surgical treatment as needed.
- d. Follow-up of patients treated for a testicular mass will vary depending on their pathology.

## Additional Notes

Acute scrotum is a general term for acute onset of pain, swelling, or tenderness to the scrotum and its contents. Although the differential diagnosis is long, there are a few emergencies to quickly rule in or out. If there is any concern for a testicular torsion, there should be no delay in proceeding with surgical intervention for emergent exploration. Often, patients are delayed due to obtaining imaging including a scrotal ultrasound for further evaluation. However, the diagnosis of testicular torsion is made clinically.

There are other reasons for delays in treatment. There are some delays in presentation due to patient embarrassment and lack of public awareness regarding torsion and its possible consequences. We hope to educate the community and primary care physicians to minimize the delays in diagnosis and inevitably, any delays in treatment due to lack of awareness.

Typically, a testicle is viable if intervened within 6–8 h of torsion. There is an inverse relationship with duration of torsion and viability of the testicle. Thus, if there is any clinical suspicion for a testicular torsion, one should proceed immediately to the operating room to increase the success of viability.

## Study Questions

1) A 10-year-old patient presents to the emergency department due to the sudden onset of left testicular pain 2 h ago. He has associated dysuria, nausea, and vomiting. His left testicle has a horizontal lie and there is an absent left cremasteric reflex. What do you do next?
   - a. Obtain a scrotal ultrasound
   - b. Obtain a urinalysis

   c. Proceed to the operating room
   d. Repeat exam

Answer to Question 1: (c.) Proceed emergently to the operating room for scrotal exploration. Testicular viability is inversely related to the duration of torsion. Thus, there should be no delays for further workup including imaging, lab work, or repeated examinations. If your clinical suspicion for testicular torsion is high, there should be no delays for surgical intervention.

2) Question 2: An 18-year-old male presents to the emergency department due to worsening right testicular pain. He reports he recently became sexually active with his girlfriend. Associated symptoms include dysuria and urinary frequency. On physical exam, patient has an intact cremasteric reflex bilaterally and a tender left testicle with normal lie. What is your likely diagnosis?

   a. Epididymitis
   b. Testicular torsion
   c. Left inguinal hernia
   d. Varicocele

Answer to Question 2: (a.) Epididymitis. Epididymitis and testicular torsion may be difficult to differentiate. Key findings that are suggestive of epididymitis include a normal lie of the testicle and intact cremasteric reflex. Since the clinical suspicion for testicular torsion is low, further workup may be performed including obtaining additional blood work, a urinalysis, and a scrotal ultrasound. For bacterial epididymitis, a course of antibiotics is warranted.

3) Question 3: The mother of an 8-year-old-male brings her son to your office for evaluation of testicular pain. She reports the pain awoke him from his sleep. She has given him pain medications and ice to the scrotum which seem to help. You notice on your exam that he has a blue discoloration of the left scrotum. What is the likely diagnosis?

   a. Hydrocele
   b. Epididymitis
   c. Testicular torsion
   d. Torsion of appendix testis

Answer to Question 3: (d.) Torsion of appendix testis. Appendage scrotum is the most common cause of acute scrotal pain in prepubertal children. Although not always present, early on, a “blue dot sign” may represent the ischemic appendage through the scrotal skin. Management is typically conservative with pain medication and ice packs. If there is a high suspicion for testicular torsion, management should be with surgical exploration.

## References

1. Partin AW, Dmochowski RR, Kavoussi LR, Peters CA, Wein AJ. Campbell-Walsh-Wein urology. 12th ed. Oxford: Elsevier; 2020.
2. Smith JA, Howards SS, Preminger GM, Dmochowski RR. Hinman's atlas of urologic surgery. 4th ed. Oxford: Elsevier; 2020.
3. Gargollo PC, Granberg CF, Lambert S (2021) Common penile and scrotal conditions. American Urological Association Core Curriculum, Pediatric Urology. AUA University. https://university.auanet.org/core/pediatric/common-pediatric-penile-and-scrotal-conditions/index.cfm, Accessed 22 Oct 2021

# Pilonidal Disease

**Savannah Rausch and Marc S. Lessin**

**Key Points**

1) Pilonidal disease is a common problem in the adolescent population. It is usually associated with entrapped hairs.
2) While there is lack of literature and research designated to the pediatric population, growing evidence supports minimally invasive measures. These include "pit picking" and placement of vessel loops/seton to be as effective as wide incisions.
3) Midline incisions should not be used. Off midline incisions are more effective to promote wound healing.
4) Occasional flap procedures are required for severe refractory disease.
5) Ongoing care and hygiene are required for successful healing. Laser ablation of hair in the sacral region has been effective in preventing recurrence.

## Background

Pilonidal disease is an inflammatory condition of the skin and subcutaneous tissues overlying the sacrum that affects primarily adolescents and young adults. The disease is more common in males. Pilonidal disease is the result of entrapped hairs in the midline natal cleft overlying the sacrum. The entrapped foreign body hair and debris leads to sinus tracts which may drain through the pit at the base of the hair

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follicle. Entrapped hairs may lead to abscesses. The hairs become ingrown as a result of shed hair that becomes burrowed into the skin due to friction. Pilonidal disease may present acutely with a painful abscess, or as a subacute or chronic infection manifest by draining pilonidal sinus tracts.

1. Epidemiology

   The incidence of pilonidal disease in the adolescent population is increasing. It is most commonly encountered in adolescents and young adults. The disease occurs about 3–4 times more frequently in males than females. It affects approximately 2.5 per 10,000. Peak incidence in the adolescent and young adult population is aged 14–25 years.

2. Pathophysiology:
   a. Anatomy: Pilonidal disease occurs in the gluteal cleft. The gluteal cleft is the indented area between the gluteus maximus muscles overlying the sacrum. Pilonidal disease should be distinguished from perirectal abscess. Pilonidal disease is not near the anus and overlies the sacrum. There may be sinus tracts known as "pits" with protruding hairs. There may be a midline or off-midline fluctuant mass.
   b. Physiology: Pilonidal disease is caused by inflammation and/or infection of the gluteal cleft and sacrococcygeal region. Typical presentation of pilonidal disease is sinus or abscess formation along the midline near the sacrum with small pits or openings. The openings contain entrapped or ingrown hair and debris. The disease can be relatively minor with small pits to large infected areas with multiple sinus tracts or fistulas away from the midline. Risk factors associated with pilonidal disease are obesity, deep gluteal cleft, hirsutism, and friction, which causes the hairs to become ingrown or embedded.
   c. Classification Schemes: Pilonidal disease may present as an acute or subacute infection or chronic condition which has been present for months to years.

3. Clinical features: Clinical feature of pilonidal disease is sinus or abscess formation near gluteal cleft with small pits or openings. The openings contain entrapped or ingrown hair and debris. Patients often report pain, bleeding, or drainage from the area. The disease may present acutely as an abscess or as a chronic condition that may be present for prolonged periods.
4. Diagnosis: The diagnosis of pilonidal disease is based on history and physical examination findings. The patient may report an acute onset or waxing and waning symptoms including drainage over months to years.
   a. Symptoms: Acute pilonidal disease presents with a painful fluctuant mass in the intergluteal area overlying the sacrum. Chronic disease presents as draining sinus tracts with a history of staining of undergarments.
   b. Examination: Physical examination is typically done with the patient lying prone or on their side. A fluctuant mass may be present in acute disease. The mass is typically off midline but tracts to the gluteal cleft. There may be over-

lying erythema and pain to palpation. Sinus tracts are often midline and may have protruding hairs or debris, which are loose. Midline pit openings represent classic pilonidal disease.

c. Radiologic studies and laboratory tests are not helpful for this condition, except in the rare circumstance in which osteomyelitis of the sacrum is suspected. The patient may present with fever and leukocytosis if an abscess is present.

5. Treatment: Optimal treatment for pilonidal disease has not been defined. Treatment can range from relatively minor disease with non-operative management to advanced disease with wide excisions and flaps.

   a. Initial Measures: Non-operative management may be considered if the pits are relatively small and asymptomatic without abscess. Non-operative management should include removal of hair or debris from the openings, hygiene, and surrounding hair removal. Removal of hair and debris from the pits requires frequent attention by the patient and the assistance of a caregiver at home. Bathing with soaking the area in warm water twice per day is recommended. Non-infected sinus tracts can be resected in an elliptical fashion with primary closure. Postoperative wound complications such as dehiscence are common. Primary closure has been shown to be superior to open wounds, particularly midline wounds.
   b. Acute Disease with Abscess: Initial treatment for pilonidal disease begins with infection control with oral antibiotics for infection including abscess if there is a delay to operation or drainage. Acute disease with an abscess requires incision and drainage, as with any abscess. Off midline incisions should be used. Midline incisions are avoided due to delayed healing. The abscess cavity should be debrided of hair, granulation tissue, and debris. Midline sinus tracts should be opened with a punch biopsy instrument. Typically a 2 or 3 mm punch biopsy instrument is used. Vessel loops in the form of seton may be placed from the sinus tracts through the incision. Packing of the wound may be indicated for hemostasis with the removal of the packing in 2–3 days. The patient is instructed to shower or bathe and keep the area clean once packing is removed. Abscess are typically polymicrobial. Typical organisms are Bacteroides, Staphylococcus, Streptococcus, Escherichia Coli, Proteus, and Pseudomonas. Postoperative antibiotics are controversial. Once the abscess is drained, antibiotics are no longer needed.
   c. Chronic Pilonidal Disease without abscess: There is rising evidence to support minimally invasive techniques as the first line operative treatment. For chronic disease with draining sinus, a minimalistic approach described by Dr. Gips has been used. A punch biopsy instrument is used to open the sinus tract. The tract is debrided of hair and debris using a curette instrument. Often multiple interventions are required with this technique. Seton may also be placed to keep the tract open for drainage. Excision of the midline sinus tracts with primary closure may be appropriate when there are multiple midline

sinus tracts. Primary closure has the advantage of faster healing when successful but is associated with higher recurrence rates than wounds healing by secondary intention.

d. Severe or Chronic Recurrence: For chronic, severe, or recurring disease, flap procedures may be necessary to obliterate the gluteal cleft. Several techniques have been described over the past 60 plus years, including the Bascom cleft lift, Limberg / rhomboid flap, Karydakis flap, V-Y flaps, and Z-plasty. These techniques allow for wide debridement, tension-free closure off midline and flattening of the gluteal cleft. No single technique has been shown to be superior (Fig. 1).
e. Preoperative Preparation: If abscess is present, prompt drainage should be the primary treatment. If there is a lag until drainage, the patient should be treated with oral antibiotics. Patient should begin good hygiene of the area leading up to surgery. This includes daily showers, avoiding prolonged periods of sitting, and hair removal from the area.
f. Postoperative Treatment: Entrapped hairs are a major contributing factor to pilonidal disease. Therefore, hair removal would make sense as part of the

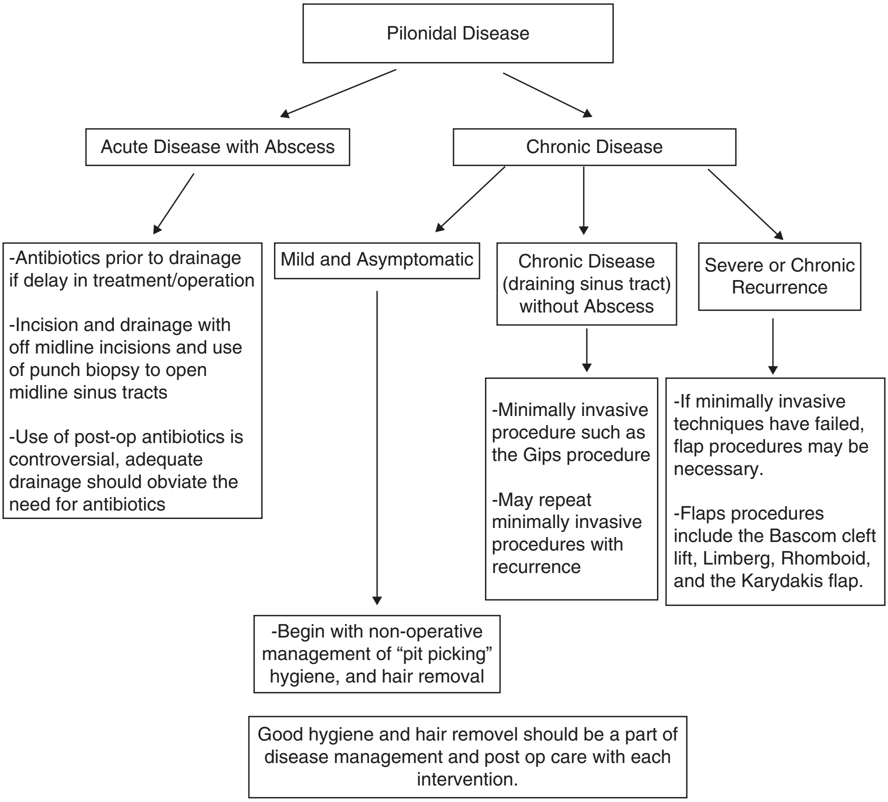
Pilonidal Disease
Acute Disease with Abscess
Chronic Disease
-Antibiotics prior to drainage if delay in treatment/operation
-Incision and drainage with off midline incisions and use of punch biopsy to open midline sinus tracts
-Use of post-op antibiotics is controversial, adequate drainage should obviate the need for antibiotics
Mild and Asymptomatic
Chronic Disease (draining sinus tract) without Abscess
Severe or Chronic Recurrence
-Minimally invasive procedure such as the Gips procedure
-May repeat minimally invasive procedures with recurrence
-If minimally invasive techniques have failed, flap procedures may be necessary.
-Flaps procedures include the Bascom cleft lift, Limberg, Rhomboid, and the Karydakis flap.
-Begin with non-operative management of "pit picking" hygiene, and hair removal
Good hygiene and hair removel should be a part of disease management and post op care with each intervention.

**Fig. 1** Algorithm for pilonidal disease (Source: Savannah Rausch, FNP)

treatment plan. However, there is no consensus on the most effective method of hair removal. Hair shaving with razors has been shown to increase risk of recurrence and infection and should be avoided. Laser hair ablation has shown decreased recurrence rates. This technique requires multiple treatments and insurance coverage is problematic due paucity of strong evidence supporting the efficacy of the method in preventing recurrence.

g. Potential Complications: While not a complication, it is important to note pilonidal disease as a whole has a high rate of recurrence. Recurrence rates occur in about 20% after complete healing. It is important that patients and families are taught proper hygiene and wound care, both pre- and post-operatively. Complete healing takes commitment and diligence by both providers and patients.

## Additional Notes

Pilonidal disease was first described by the famous surgeon Herbert Mayo in the nineteenth century. It was originally thought to be a congenital condition but is now known to be an acquired condition. As the term implies from its Latin roots of pilus meaning hair and nidal meaning nest; the condition arises from ingrown hairs in the skin overlying the sacrum. It has been referred to as pilonidal cyst but this is a misnomer since it lacks an epithelial lining. It was often described after World War II as “jeep driver’s disease” because of the propensity of soldiers to acquire this condition. It is a major source of disability for military personnel in the past and present. The disease is commonly seen in those with deep gluteal cleft, hirsutism, obesity, and friction which causes the hairs to become impaled in the gluteal region. Traditional techniques of wide local excision with primary or delayed closure done in the past are no longer standard of care. Midline incisions need to be avoided whenever possible as they are associated with increased recurrence and complication.

Lord and Millar first suggested a less invasive approach with limited excision of the involved area and debridement. Bascom described the technique of lateral drainage and pit picking. There is a current resurgence toward these less invasive techniques which have been shown to have similar recurrence rates with less morbidity associated with open wounds.

Flap procedures are done in some cases as primary procedures but more commonly for chronic and recurrent diseases. Flap procedures were first introduced by Limberg in 1946 to close midline wounds using a full-thickness rhomboid-shaped rotational flap which flattens the gluteal cleft. Karydakis described an advancement flap in using an off-midline incision on the more affected side with excision of the diseased skin. The flap is secured in layers with a deep layer to the sacrococcygeal fascia. Karydakis reported recurrence rates of less than 2% in over 6500 patients using the flap procedure that bears his name. Bascom and Bascom described the cleft-lift technique which involves excision of an ellipse of the diseased skin with advancement across the midline decreasing the depth of the gluteal cleft. A 7-mm flap is raised opposite the elliptical incised tissue and advanced across the midline with primary closure. Abscess walls are left removing granulation tissue from the surfaces of cavities and tunnels. The difference in this technique from Karydakis's technique is that the flaps are not full thickness and the underlying tissue including the fat and cavity wall are not excised.

Dr. Gips described a variation to the original Bascom pit-picking procedure in which skin trephines (cylindrical excision typically using an instrument similar to a punch biopsy but able to penetrate deeper into the tissue by lacking the shelf at 8 mm) are used to core out all tracts. Curettes and cotton swabs with hydrogen peroxide were placed through the opening to remove hair, debris, and granulation tissue leaving the wounds open and unpacked to heal by secondary intention. The procedure can be repeated if necessary. The authors report recurrence rates associated with this technique of 6.5% at 1 year, 13.2% at 5 years, and 16.2% at 10 years. We have modified this technique by placing vessels loops as setons.

At present, the most optimal management needs to be determined on a case-by-case basis. Minimal invasive techniques should be the initial management with avoidance of large open incisions. When incisions are necessary, off-midline incisions with circular skin incisions of midline sinuses should be the first modality. Large midline incisions should be avoided. For recurrent disease, flap procedures should be considered.

## Study Questions

1. Sixteen-year-old male presents with chronic drainage on the underwear. Physical examinations show several sinus tracts in the midline with protruding hairs. What is the most appropriate treatment?

   a. Wide elliptical incision with primary closure.
   b. Wide elliptical incision with open wound and packing
   c. Punch biopsy of the opening with debridement of hair follicles
   d. Off midline incision with debridement from lateral to medial

Answer to Question 1: Option C. Several of the options may be acceptable but a minimally invasive approach for the primary procedure is the most appropriate choice. An off- midline incision with primary closure has been shown to effectuate better healing than midline incisions. The Gips procedure described by Dr. Gips as opposed to an elliptical midline incision with or without open wounds has been shown to have similar recurrence rates, faster healing with earlier return to activity, and less morbidity.

2. 17-year-old female presents with one-week history of acute onset of pain over the pilonidal region. The patient has a fever of 101° and complains of an inability

to sit. On exam, there is erythema overlying the sacrum with a fluctuant mass extending to the left of midline. There are 2 midline sinus tracts with hairs protruding. The patient is morbidly obese with a BMI=36 kg/m$^2$. The patient has significant hair growth in the gluteal area and a deep gluteal cleft. The most appropriate initial treatment is?

a. Bedside incision and drainage midline with oral antibiotics.
b. Operative drainage off-midline with opening of the pilonidal sinus tracts with debridement and vessel loops (as Seton) from the incision to one or both of the pilonidal sinus tracts.
c. Operative drainage off midline with packing of the wound.
d. Midline elliptical incision of the sinus tracts with debridement of the abscess cavity with open wound and packing.

Answer to Question 2: B. Patient presenting with a pilonidal abscess requires drainage and debridement as would be required for any abscess. Midline incisions should be avoided. It is important to open the midline pilonidal sinus tracts and debride both from the lateral incision and through the sinus tracts. Packing may be placed in the off midline wound but should be removed in 2–3 days. Seton suture can be used for long-term drainage.

3. You have just performed a minimally invasive Gips procedure on a 15-year-old male for chronic pilonidal disease. His family is asking for post-op instructions. Which of the following is the most appropriate answer?

   a. Shower daily. Remove hair by shaving the gluteal cleft once per week. Resume normal physical activity as tolerated.
   b. Bathe and remove any entrapped hair daily. Shave the gluteal cleft once per week or undergo laser hair ablation. Activity restrictions should be in place for 2–4 weeks with limited daily physical activity.
   c. Shower or bathe one to two times per day. Hair and debris from the pilonidal pits should be removed multiple times per week if someone at home is able to assist. Shave the gluteal cleft once per week but do not use a razor. May resume normal physical activity as tolerated and avoid sitting for prolonged periods of time.
   d. Shower or bathe one to two times per day. For infection, treat with antibiotics for 7 days. May resume normal physical activity as tolerated and avoid sitting for prolonged periods of time.

Answer to Question 3: C. Patients should be encouraged to shower/bathe at minimum once per day. Hair removal is important to prevent recurrence and allow for healing. However, razor shaving should be avoided as it is associated with increased recurrence. If the patient is awaiting either an appointment for or insurance authorization for laser hair ablation, hair removal products can be used in the meantime. Hair and debris prevents the pilonidal sinus tracts and pits from healing. It is important to remove hair/debris frequently. It should be done multiple times per week and waiting for follow-up appointments will not be adequate. Family can be taught to remove debris with fine tweezers. There is no need for physical activity restrictions within reason and as patient tolerates. Prolonged sitting or immobilization should be avoided. Antibiotics typically do not play a role in chronic disease. If there is no active infection and the site has been adequately debrided, no post-op antibiotics are indicated.

## Further Reading

Al-Khamis A, McCallum I, King PM, Bruce J. Healing by primary versus secondary intention after surgical treatment for pilonidal sinus. Cochrane Database Syst Rev. 2010;1:CD006213.

Bascom J. Pilonidal disease: origin from follicles of hairs and results of follicle removal as treatment. Surgery. 1980;87(5):567–72.

Bascom J, Bascom T. Failed pilonidal surgery: new paradigm and new operation leading to cures. Arch Surg. 2002;137(10):1146–50.

Delshad HR, Dawson M, Melvin P, et al. Pit-picking resolves pilonidal disease in adolescents. J Pediatr Surg. 2019;54(1):174–6.

Di Castro A, Guerra F, Sandri GBL, Etorre GM. Minimally invasive surgery for the treatment of pilonidal disease. The Gips procedure on 2347 patients. Int J Surg. 2016;36(Part A):201–5.

Gips M, Melki Y, Salem L, et al. Minimal surgery for pilonidal disease using trephines; description of a new technique and long-term outcomes in 1,358 patients. Dis Colon Rectum. 2008;51(11):1656–63.

Grabowski J, Oyetunji TA, Goldin AB, et al. The management of pilonidal disease: a systematic review. J Pediatr Surg. 2019;54(11):2210–1.

Halleran DR, Onwuka AJ, Lawrence AE, et al. Laser depilation in the treatment of pilonidal disease: a systematic review. Surg Infect (Larchmont). 2018;19(6):566–72.

Karydakis G. Easy and successful treatment of pilonidal sinus after explanation of its causative process. ANZ J Surg. 1992;62(5):385–9.

Millar DM, Lord PH. The treatment of acute postanal pilonidal abscess. Br J Surg. 1965;54:598–9.

# Metabolic and Bariatric Surgery in Pediatrics

**Max D. Hazeltine and Jeremy T. Aidlen**

**Key Points**

1) Obesity is an epidemic in the adolescent population. Surgery for obesity provides the best long-term outcomes for sustained weight loss and resolution of comorbidities.
2) Indications for metabolic and bariatric surgery include (A) BMI ≥35 kg/m$^2$ or 120% of the 95th percentile with a clinically significant comorbid condition, or (B) BMI ≥40 kg/m$^2$ or 140% of the 95th percentile (whichever is lower), in addition to readiness for change, and the ability of the patient and family to adhere to treatments pre- and postoperatively.
3) Vertical sleeve gastrectomy is the most common weight loss procedure performed in adolescents. Roux-en-Y gastric bypass may be preferred in patients with severe gastroesophageal reflux disease.
4) A dedicated adolescent bariatric surgery program with lifelong monitoring postoperatively is essential for optimal patient outcomes.

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Obesity is the single most prevalent chronic disease in childhood and adolescence. It is a complex, progressive multi-metabolic and hormonal condition with few successful treatment options. The treatment of pediatric patients with obesity has evolved significantly over the past two decades. Bariatric surgery, once reserved only for adults, has been shown to reliably improve body mass index (BMI), comorbidity burden, and quality of life for pediatric patients in large controlled trials [1].

1. Epidemiology
    a. Approximately 20% of youth in the USA meet the criteria for obesity (body mass index [BMI] percentile ≥95th for age and sex), and 8.5% of adolescents aged 12 to 19 years are categorized as severely obese (BMI ≥120% of the 95th percentile or ≥35 kg/m$^2$). Severe obesity is the fastest-growing subcategory of obesity.
2. Pathophysiology
    a. Obesity is a multifactorial disease, caused by a combination of genetic and environmental factors as well as other comorbid conditions.
    b. Up to 200 types of single-gene mutations have been found to cause obesity. A few well-known monogenic mutations cause <10% of cases of severe obesity, such as leptin or leptin receptor mutations. A small proportion of cases are related to syndromes such as Prader-Willi. Epigenetic changes, which are changes in DNA modification and expression that are susceptible to lifestyle changes that may be heritable, are implicated in the development of obesity.
    c. Environmental factors include less physical activity, increased caloric intake, and use of medications with weight gain as a side effect.
    d. Comorbidities that have a role in weight regulation include diabetes, hypothyroidism, Cushing disease, pseudohypoparathyroidism, growth hormone deficiency, hypothalamic disorders, and polycystic ovary syndrome.
3. Classification
    a. Overweight: BMI ≥85th percentile to <95th percentile
    b. Class I obesity: BMI ≥30 kg/m$^2$ to 34.9 kg/m$^2$ and/or BMI ≥95th percentile to <120% of the 95th percentile
    c. Class II obesity (severe): BMI ≥120% to <140% of the 95th percentile or a BMI ≥35 to ≤39.9 kg/m$^2$
    d. Class III obesity (severe): BMI ≥140% of the 95th percentile or BMI ≥40 kg/m$^2$
4. Obesity-Related Comorbidities
    a. Cardiovascular disease
        i. Obesity is associated with the development of numerous cardiovascular disease (CVD) risk factors such as hyperlipidemia, elevated inflammatory markers, hypertension, and insulin resistance.

    ii. Children with obesity have a three–five times higher risk of CVD-related mortality at age 50 years.
    iii. There are significant improvements in CVD risk factors by 2 years after metabolic and bariatric surgery (MBS).
    iv. Younger patients may be more likely to have improvements in cardiovascular risk factors compared with older patients.

b. Type 2 diabetes

    i. Childhood obesity associated with increased incidence of type 2 diabetes (T2D).
    ii. T2D is often preceded by insulin resistance.
    iii. Childhood-onset T2D is associated with earlier end organ injury and more medically refractory T2D.
    iv. Weight loss surgery produces remission of insulin resistance and T2D in approximately 90% of adolescents.

c. Obstructive sleep apnea

    i. Approximately one-third of children and adolescents with obesity have obstructive sleep apnea (OSA).
    ii. Children with OSA have increased rates of morbidity and mortality.
    iii. Obstructive sleep apnea is associated with increased right ventricular pressure leading to pulmonary hypertension.
    iv. Weight loss surgery leads to significant improvement or resolution of OSA.

d. Nonalcoholic fatty liver disease and steatohepatitis

    i. Of those undergoing bariatric surgery, nonalcoholic fatty liver disease (NAFLD) present in upwards of two-thirds of patients.
    ii. Weight loss is the primary method to manage NAFLD.
    iii. Long-term sequelae of NAFLD include cirrhosis and liver failure.
    iv. Weight loss surgery leads to nonalcoholic steatohepatitis (NASH) resolution in 85% of patients, and one-third will have improvement in fibrosis.

e. Idiopathic intracranial hypertension

    i. Idiopathic intracranial hypertension (IIH) is defined as elevated intracranial pressure without a cause determined by clinical, radiologic, or laboratory studies.
    ii. IIH is also known as pseudotumor cerebri or benign intracranial hypertension.
    iii. IIH can lead to permanent visual impairment or blindness.
    iv. There is a strong association between childhood obesity and IIH risk.
    v. Weight loss has been shown to reduce intracranial pressure.

f. Orthopedic disease

    i. Pediatric specific diseases that occur due to childhood obesity include Blount's disease (tibia vara) and slipped capital femoral epiphysis.

    ii. Metabolic bariatric surgery improves functional mobility.
    iii. Metabolic bariatric surgery may improve orthopedic outcomes and reduce the risk of developing bilateral or recurrent disease.

g. Gastroesophageal reflux disease
    i. Gastroesophageal reflux disease (GERD) is often associated with obesity and is likely exacerbated by increased intraabdominal pressures.
    ii. Weight loss improves GERD.
    iii. Roux-en-Y gastric bypass (RYGB) is highly effective in treating GERD and is the surgical treatment of choice for patients with obesity and severe GERD.
    iv. GERD may occur after vertical sleeve gastrectomy (VSG) in upwards of 60% of patients, although weight loss can reduce preexisting GERD.

h. Mental health
    i. Behavioral disorders, especially depression, are commonly associated with obesity.
    ii. Psychopathology may be a barrier to approval for weight loss surgery, and is also predictive of the inability to complete the preoperative phase prior to surgery.
    iii. Metabolic bariatric surgery likely leads to improved mental health outcomes (i.e., depressive symptoms or disruptive behaviors).
    iv. Substance use disorders are also significant factors, with alcohol being the most abused substance. Alcohol use increases after weight loss surgery, likely as a byproduct of increasing age.
    v. All patients should be screened for alcohol use and should also be discouraged from smoking as both are associated with ulcer formation.
    vi. Disordered eating occurs in approximately one in four patients undergoing metabolic bariatric surgery and is associated with poor outcomes postoperatively. However, disordered eating may be related to other psychopathology which is treatable.
    vii. Identification of disordered eating such as binge or purging behavior is critical preoperatively. Once identified these behaviors should be treated and found to be in remission prior to proceeding with surgery.
    viii. Symptoms that are well managed and monitored should not be considered a contraindication to weight loss surgery.
    ix. Active psychosis, suicidality, or substance abuse are contraindications.

i. Quality of life
    i. Adolescents with severe obesity report worse quality of life, such as weight-related physical comfort, self-esteem, or social life.
    ii. Weight loss surgery leads to significant long-term improvements in the quality of life for adolescents with severe obesity.

j. Family factors
    i. Obesity is often a multigenerational disease, with caregivers of children with obesity also suffering from obesity.
    ii. Family dysfunction occurs in up to half of families with a child that undergoes surgery for obesity.
    iii. However, family dysfunction does not appear to be related to poor short-term outcomes.

5. Treatment
    a. Metabolic and bariatric surgery (MBS)
        i. MBS is a proven, effective treatment for severe obesity disease in adolescents and should be considered standard of care.
        ii. Indications
            1. BMI ≥35 kg/m$^2$ or 120% of the 95th percentile with a comorbidity.
            2. BMI ≥40 kg/m$^2$ or 140% of the 95th percentile (whichever is lower).
            3. Patient and family are able to adhere to treatments pre- and postoperatively, including use of micronutrient supplements, as determined by a multidisciplinary team.
        iii. Contraindications
            1. Medically correctable cause of obesity.
            2. Substance abuse problem within the last year.
            3. Medical, psychiatric, psychosocial, or cognitive condition that prevents adherence to postoperative dietary and medication regimens.
            4. Current or planned pregnancy within 12 to 18 months of procedure.
            5. Puberty and linear growth status do not impact eligibility.
    b. Preoperative Preparation
        i. MBS programs require a multidisciplinary team, including a pediatrician, dietitian, psychologist, psychiatrist, or other behavioral specialist, and a surgeon to evaluate the patient and family.
        ii. Patient must demonstrate the ability to make lifestyle changes that will be required postoperatively to maintain weight loss and healthy habits.
    c. Operative Treatment
        i. Vertical sleeve gastrectomy (VSG). This is currently the most common MBS performed for pediatric patients. The VSG removes the entire greater curve of the stomach (~80% of the stomach) using a linear stapling technique. The remaining stomach has a volume of 60–100 ml. This restrictive procedure has slightly lower weight loss than rouxen-Y gastric bypass (RYGB) but has approximately one-third the rate of serious complications.

    ii. Roux-en-Y gastric bypass. The RYGB involves creating a small gastric pouch that is anastomosed to a jejunal limb that bypasses the gastric remnant as well as duodenum and proximal jejunum. This procedure offers restriction and malabsorption and has been shown to be superior to VSG for the treatment of GERD and leads to slightly greater weight loss. However, it should be avoided in patients who have issues with medication compliance or may be at risk for ulcer formation postoperatively (smoking, living with those who smoke).
    iii. Adjustable gastric banding. This procedure is currently not FDA approved in patients <18 years of age unless within the context of a clinical trial. This procedure is also associated with an over 40% reoperation rate. It is not recommended in the adolescent population and has largely been replaced by vertical sleeve gastrectomy in the adult population.
    iv. Biliopancreatic diversion with or without duodenal switch. This malabsorptive procedure is not recommended in adolescents as it can lead to significant morbidity postoperatively.

d. Postoperative Treatment

    i. Lifestyle modification—A commitment to healthy behavioral change preoperatively should continue after surgery. This may include avoiding triggers to disordered eating, for example.
    ii. Dietary recommendations—Hydration and liquid protein calories are the mainstay of the postoperative diet for weeks prior to the gradual reintroduction of solid food.
    iii. Exercise—A gradual and achievable exercise program is helpful for motivation and strengthening postoperatively. Maintenance of muscle mass is important for healthy weight loss.
    iv. Lifelong vitamin level monitoring and supplemental vitamins—commonly includes a multivitamin, calcium, B12, and iron supplementation, but this varies by patient and program.
    v. Medical treatment for comorbidities.
    vi. Patients may eventually need to transition to an adult metabolic and bariatric program.
    vii. RYGB patients require lifelong gastric acid suppression.

e. Complications

    i. RYGB specific—marginal ulcers, internal hernias
    ii. VSG specific—GERD
    iii. Bleeding
    iv. Anastomotic or staple line leaks
    v. Strictures
    vi. Failure to lose weight
    vii. Weight regain
    viii. Nutritional deficiencies are common in all procedures including VSG (most commonly hypoferritinemia).

ix. Anemia
x. Thromboembolic events
xi. Death is rare (<1%)

Obesity disproportionally affects minorities and those with socioeconomic disadvantage. As pediatric bariatric surgery programs become more prevalent, it will be important to recognize and address known disparities in access to care.

The oversight, so far, in the treatment of pediatric obesity has been a failure to recognize this disease as a killer. Long-term exposure to obesity can lead to a gradual progression of cardiometabolic disease and early death. Teens with BMI >99th percentile are always going to suffer from obesity as adults.

While prevention should remain the mainstay of treatment, patients struggling with a high BMI and comorbidities have improved outcomes with early referral and supportive discussion about the dangers of obesity and benefits of surgical intervention. The treatment of pediatric obesity requires a multidisciplinary approach, and bariatric surgery should not be considered a last resort [2]. Timeliness of referral is crucial as long-term exposure to excess weight compounds, comorbid conditions, leading to a higher preoperative, and higher postoperative weight. Alternative modalities of treatment currently available to pediatric patients all fall short in comparison to surgery.

Vertical sleeve gastrectomy has surpassed RYGB to become the most commonly performed bariatric surgical procedure for pediatric patients. Adjustable gastric band surgery was never FDA approved for patients under the age of 18 years and has fallen out of favor due to inferiority of comorbidity resolution and a high secondary intervention rate. While sleeve gastrectomy is less malabsorptive than RYGB, nutritional deficiencies are still common and lifelong micronutrient supplementation and monitoring is necessary. A comprehensive team approach including behavioral, nutritional, medical, and surgical support is a prerequisite to long-term success, as is patient compliance and readiness for change.

## Study Questions

1) Question 1: A 17-year-old male with a history of severe GERD presents for MBS evaluation. He is deemed a surgical candidate. Which of the following procedures would be preferred?

   a. Vertical sleeve gastrectomy
   b. Roux-en-Y gastric bypass
   c. Adjustable gastric banding
   d. Biliopancreatic diversion without duodenal switch

Answer to Question 1: B-Roux-en-Y gastric bypass, as this is the optimal treatment for a patient with concomitant GERD. Vertical sleeve gastrectomy can lead to worse GERD symptoms postoperatively.

2) Question 2: A 15-year-old female undergoes a VSG. The case is uneventful, and she recovers well overnight. The following day you see her on rounds and note that her heart rate is 115, which was previously between 80 and 90, and has a temperature of 38.4 degrees Celsius. On physical exam, she is more tender in the left upper quadrant than when she was in the recovery room. What is the primary concern given your findings?

   a. Staple line leak
   b. Staple line bleeding
   c. Pulmonary embolus
   d. Urinary tract infection
   e. Surgical site infection

Answer to Question 2: A-This patient is presenting with signs and symptoms of a staple line leak given progressive tachycardia, fever, and worsening tenderness on exam. In addition to initiation of antibiotics, the appropriate management would be re-exploration, identification of the source of the leak, and repair.

3) Question 3: A 16-year-old male underwent a RYGB one year ago with a resultant decrease in BMI by 15 kg/m$^2$. He has been compliant with his postoperative vitamin regimen, takes no other medications, and has been to all of his follow-up visits. He develops acute onset abdominal pain with associated nausea and vomiting. What is the most likely diagnosis?

   a. Gastroenteritis
   b. Marginal ulcer
   c. Anastomotic leak
   d. Internal hernia

Answer to Question 3: D-This patient has had significant weight loss postoperatively, has been adherent to his vitamin regimen, and is not taking any medication to increase the risk of ulcer formation (NSAIDs). Given the timeframe (one year postoperatively), weight loss, and adherence to the vitamin regimen, he most likely has an internal hernia. Internal hernias most often occur after a significant amount of weight loss, are more common after RYGB, and clinically present like a small bowel obstruction (nausea, vomiting, abdominal pain, and distention). These are surgical emergencies that warrant operative exploration.

## Bibliography

1. Inge TH, Courcoulas AP, Jenkins TM, Michalsky MP, Helmrath MA, Brandt ML, et al. Weight loss and health status 3 years after bariatric surgery in adolescents. N Engl J Med. 2016;374(2):113–23.
2. Pratt JSA, Browne A, Browne NT, Bruzoni M, Cohen M, Desai A, et al. ASMBS pediatric metabolic and bariatric surgery guidelines, 2018. Surg Obes Relat Dis. 2018;14(7):882–901.

3. Ogden CL, Carroll MD, Fryar CD, Flegal KM. Prevalence of obesity among adults and youth: United States, 2011–2014. NCHS Data Brief. 2015;219:1–8.
4. Skinner AC, Ravanbakht SN, Skelton JA, Perrin EM, Armstrong SC. Prevalence of obesity and severe obesity in US Children, 1999–2016. Pediatrics. 2018;141(3):e20173459.
5. Kelly AS, Barlow SE, Rao G, Inge TH, Hayman LL, Steinberger J, et al. Severe obesity in children and adolescents: identification, associated health risks, and treatment approaches: a scientific statement from the American Heart Association. Circulation. 2013;128(15):1689–712.
6. Upadhyay J, Farr O, Perakakis N, Ghaly W, Mantzoros C. Obesity as a disease. Med Clin North Am. 2018;102(1):13–33.
7. Skinner AC, Skelton JA. Prevalence and trends in obesity and severe obesity among children in the United States, 1999–2012. JAMA Pediatr. 2014;168(6):561–6.
8. Zeller MH, Hunsaker S, Mikhail C, Reiter-Purtill J, McCullough MB, Garland B, et al. Family factors that characterize adolescents with severe obesity and their role in weight loss surgery outcomes. Obesity (Silver Spring). 2016;24(12):2562–9.
9. Bolling CF, Armstrong SC, Reichard KW, Michalsky MP. Metabolic and bariatric surgery for pediatric patients with severe obesity. Pediatrics. 2019;144(6):e20193224.
10. Peña AS, Delko T, Couper R, Sutton K, Kritas S, Omari T, et al. Laparoscopic adjustable gastric banding in Australian adolescents: should it be done? Obes Surg. 2017;27(7):1667–73.

# Part VIII
# Common Neoplasms in Children and Adolescents

# Wilms Tumor

**Jagadeesh Ramdas**

**Key Points**

1) Identify the Wilms tumor predisposing syndromes
2) Significance of proper staging and treatments
3) Understand the role of pre-nephrectomy chemotherapy especially in bilateral Wilms tumor

1. Epidemiology
    a. Incidence: Wilms tumor represents about 6% of childhood cancer, with a total incidence in the USA of about 500 cases per year and represents over 90% of all childhood renal tumors. Most commonly seen before 5 years of age. It is the second most common solid abdominal tumor in children (neuroblastoma most common).
    b. Other renal tumors:
        1. Rhabdoid tumors, 1 %: primarily in infants and have brain metastasis.
        2. Clear cell sarcoma of kidney, 2 %: usually below 5 years old and have both bony and CNS metastatic disease.
        3. Renal cell carcinoma: Most common in adults, only 2 % occur in patients below 20 years old.
        4. Mesoblastic nephroma, 2%: Most common renal tumor of the neonatal period. It may be malignant in approximately 10% of cases.

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2. Etiology: Wilms tumor can occur as part of several rare congenital syndromes.
   a. WAGR syndrome: Wilms tumor, aniridia, genitourinary malformation, mental retardation.
   b. Denys-Drash Syndrome: Male disorders of sexual development, degenerative renal disease (glomerulonephritis or nephrotic syndrome) with Wilms tumor.
   c. Beckwith-Wiedemann Syndrome- Macroglossia, omphalocele, visceromegaly, with or without hemihypertrophy. Other tumors associated with Beckwith-Wiedemann Syndrome are hepatoblastoma, rhabdomyosarcoma, and adrenal cortical carcinoma.
   d. Simpson-Golabi-Behmel syndrome: Overgrowth syndrome resulting from deletion of the GPC3 gene.
   e. No consistent correlation between any environmental exposures for Wilms tumor patients or their parents has been identified.

3. Pathology:
   a. Most Wilms tumors are unilateral lesions. 7% are bilateral and 12% are multifocal.
   b. Wilms tumors are usually sharply demarcated-spherical masses with a pushing border related to the renal parenchyma and usually form a fibrous pseudocapsule.
   c. Microscopically the classic nephroblastoma consists of varying proportions of three cell types including blastemal, stromal, and epithelial. Not all specimens are triphasic and can present as monophasic Wilms tumor.
   d. Histology:
      i. Favorable histology are well differentiated tumors that resemble developing embryonic renal tissue with the presence of all three histologic features: tubules, blastema, and stroma.
      ii. Unfavorable histology: anaplastic Wilms tumor—anaplasia may focal or diffuse.
      iii. Nephrogenic rests are precursor lesions to Wilms tumor. They are composed of abnormally persistent embryonal nephroblastic tissue with small clusters of blastemal cells, tubules, or stromal cells. They can be either intralobar or perilobar nephrogenic rests. The term “nephroblastomatosis” is used to refer to the presence of multiple nephrogenic rests. Hyperplastic nephrogenic rests may completely regress or differentiate following chemotherapy administration.
      iv. Nephrogenic rests in nephrectomy specimens identifies those at risk of developing additional Wilms tumor in the contralateral kidney requiring close follow-up.

4. Anatomy
   a. The kidneys are retroperitoneal organs which lie below the diaphragm. The superior poles of the kidney are roughly posterior to the spleen on the left and the liver on the right. The adrenal glands lie superior and medial to the kidneys.
   b. The kidneys are covered by a thin fascia known as Gerota's fascia.
   c. Blood supply
      i. The arterial supply is by the renal artery which are branches of the aorta.
      ii. The renal artery divides into segmental branches. Duplicate renal arteries are sometimes present.
      iii. The right renal vein enters the inferior vena cava at a lower point than the left renal vein. It has no significant tributary branches.
      iv. The left renal vein drains into the vena cava and is longer than the right as the inferior vena cava is to the right of midline. It passes anterior to the aorta. The left gonadal vein, left adrenal vein, and left inferior phrenic vein drain into the left renal vein.
      v. The ureter extends from the hilum of the kidney along the retroperitoneum along the psoas muscle into the bladder trigone posteriorly.
5. Clinical Presentation
   a. Usual presentation is asymptomatic abdominal mass detected by family or physician.
   b. Pain, gross or microscopic hematuria in 20–30 %. May occur after minor trauma or fall.
   c. Hypertension (from distortion of renal vasculature).
   d. Anemia (from bleeding into tumor).
   e. A persistent varicocele when supine may be associated with the presence of a tumor thrombus in the renal vein obstructing flow in the gonadal vein (left-sided).
      i. Physical exam: Vital signs including blood pressure and examine for congenital abnormalities: hemihypertrophy, genitourinary anomalies, aniridia, etc.
      ii. Labs: CBC, UA, BUN, creatinine, liver function tests, and urine metanephrines to differentiate from neuroblastoma.
6. Imaging studies: Essential part of initial staging to establish the presence of a renal tumor and a normally functioning contralateral kidney, assess the contralateral kidney for tumor, to document the patency of the IVC, and to assess the lungs for metastatic disease.
   a. Abdominal ultrasound exam is usually performed first which can evaluate the renal vein for tumor thrombus.

   b. Contrast-enhanced CT of the abdomen or MRI abdomen is used to evaluate tumor invasion into adjacent structures. Careful attention should be paid to the contralateral kidney for focal abnormalities that may be nephrogenic rests or Wilms tumor.
   c. Presence of bilateral lesions will alter the surgical and therapeutic approach.
      i. Patency of the IVC needs to be demonstrated prior to surgical approach. When a tumor is identified within the IVC, proximal extent of the thrombus must be established prior to surgery because of the risk of dislodgment of unsuspected intravascular tumor thrombus.
      ii. Chest CT scan should be obtained to identify pulmonary metastases.
   d. Cytogenetic profile: deletion of short arm of chromosome 11

7. Wilms Tumor Staging:
   a. Stage I: Tumor limited to the kidney and completely excised. Renal capsule has an intact outer surface. Tumor is not biopsied or ruptured prior to removal.
   b. Stage II: Tumor extends beyond the kidney but is completely excised. No evidence of tumor at or beyond the margins of resection. Regional lymph nodes may be involved.
   c. Stage III: Residual non-hematogenous tumor confined to abdomen. Including unresectable or incompletely resected primary tumor; lymph node metastases to the para-aortic nodes, positive surgical margins, biopsy, or tumor spillage before or during surgery, or transected tumor thrombus.
   d. Stage IV: Hematogenous metastasis: Lung, liver, bone, or brain or lymph node metastasis outside the abdominopelvic region
   e. Stage V: Bilateral renal involvement at diagnosis.

8. Treatment
   a. Surgery
      i. Surgical resection is the initial treatment for most children with Wilms tumor.
      ii. North American practice is initial nephrectomy because treatments can best be modulated in their intensity according to accurate histopathological and staging criteria. (In Europe, chemotherapy is often initiated without biopsy based on imaging to shrink the tumor preoperatively).
      iii. Tumor is removed by radical or modified radical nephrectomy using a transperitoneal approach to the tumor to allow inspection of the regional lymph nodes.
      iv. Thorough exploration of the abdominal cavity is carried out to assess liver, regional lymph nodes and other evidence of tumor spread.
      v. Tenets of surgery include avoidance of spillage and biopsy of lymph nodes along the renal hilum and the para-aortic lymph nodes. Tumor

    thrombus should be removed from the renal vein and adjacent vena cava. The ureter should be excised as low as possible (to the level of the bladder) in continuity with the kidney. The adrenal gland should be salvaged if possible.

    vi. It is not necessary to mobilize and inspect the contralateral kidney if there is no evidence of lesions on CT or MRI.
    vii. Contralateral lesions identified on CT should be biopsied to determine histology as there may be metachronous lesions (favorable on one side and unfavorable on the opposite side).
    viii. Nephrectomy should be deferred, and chemotherapy used first, if complete tumor removal would require excision of all or parts of the adjoining organs that might be invaded. Inoperability should be decided at initial surgery.
    ix. Attempts at primary surgical excision of the tumor with vena-caval extension at or above the liver should not be undertaken and prenephrectomy chemotherapy given.
    x. Preoperative chemotherapy is advisable in children with massive tumors that are judged to pose too great risk for surgical removal.
    xi. For unresectable tumors, children should still undergo initial exploration to establish the diagnosis by biopsy and to assess operability.
    xii. Definitive resection should be completed once there is an adequate reduction in the size of the tumor usually at week 6 or week 12.
    xiii. Treatment of children with bilateral tumors must be individualized. Primary excision of the tumor should not be attempted, but rather patients should be given preoperative chemotherapy to maximize the possibility of successful partial nephrectomy.

b. Radiotherapy

    i. Wilms Tumor is a very radiosensitive tumor. Flank irradiation is currently used for stage III Wilms tumor along with 3-drug chemotherapy.
    ii. Abdominal irradiation is used for all stages of anaplastic Wilms tumor.
    iii. Irradiation should be delivered within 14 days of nephrectomy.
    iv. Whole lung irradiation is given for children with pulmonary metastases.

c. Chemotherapy

    i. Vincristine, dactinomycin, and doxorubicin are the three drugs that are the backbone agents for Wilms tumor chemotherapy. Patients with stage I and stage II get two drug regimens (vincristine and dactinomycin). Those with more extensive disease stage III or stage IV at diagnosis or whose tumors failed to respond adequately have been treated with doxorubicin in addition to vincristine and dactinomycin.

9. Outcomes: Wilms tumor is usually a curable malignancy; it is essential to limit any iatrogenic sequelae.
    i. Overall survival for stage I- stage IV ranges from 97%- 86%, respectively.
    ii. Long-term outcomes of Wilms tumor survivors remain good, however, there is a long-term risk of renal dysfunction. Patients have been described with proteinuria and hypertension developing 10 to 20 years after nephrectomy.
    iii. Single kidney health should be emphasized with adequate hydration, avoidance of nephrotoxic drugs, avoidance of sports or activity-related trauma to the single kidney and timely treatment of any urinary tract infection to avoid ascending pyelonephritis.
    iv. Patients with bilateral disease remain at higher risk for renal failure and will need to be more closely followed.
    v. In children with any of the Wilms tumor predisposing syndromes or with nephrogenic rests in one or both kidneys should undergo serial ultrasounds for several years as the opposite kidney continues to be at risk for developing Wilms tumor for several years.

## Additional Notes

National Wilms Study Group (NWTSG) in North America and SIOP in Europe use similar chemotherapy regimens, however, treatment approaches are fundamentally different in that NWTSG recommends initial nephrectomy, whereas SIOP advocates preoperative chemotherapy. Both approaches yield excellent outcomes for children with favorable histology Wilms tumor. Initial nephrectomy allows for greater consideration of surgical, pathologic, and biologic features in determining patient-specific therapy, whereas the post-chemotherapy nephrectomy allows the response of the tumor to chemotherapy to be considered in determining subsequent therapy.

## Study Questions

1. All the following are considered as Wilms tumor predisposing syndromes except
    a. WAGR syndrome
    b. Denys-Drash Syndrome
    c. Noonan Syndrome
    d. Beckwith-Wiedemann Syndrome

Answer to Question 1: (c.) Noonan Syndrome. WAGR syndrome: Wilms tumor, aniridia, genitourinary malformation, mental deficiency. Denys-Drash Syndrome: pseudo-hermaphroditism, degenerative renal disease (glomerulonephritis or nephrotic syndrome), Wilms tumor; Beckwith-Wiedemann Syndrome: macroglossia, omphalocele, visceromegaly, with or without hemihypertrophy. Patients with these Wilms tumor predisposing syndromes need to be closely followed with serial ultrasound and chest x-rays every 3–6 months until they are at least 8years of age.

Question 2: Abdominal irradiation after surgery is not indicated for which type of patient?

a. Stage II favorable histology
b. Patient who had initial biopsy only
c. Patient with anaplastic histology
d. Patient who had tumor spillage during surgery

Answer to Question 2: (a.) Stage II favorable histology. Abdominal irradiation can be flank (10.5 Gy) or whole abdomen. Current indication for abdominal irradiation is for patients with focal or diffuse anaplasia histology any stages, stage III, and stage IV favorable histology. Whole abdomen is usually given in the event of positive lymph nodes and gross tumor spillage

Question 3: Which of the following is an acceptable treatment option for bilateral Wilms tumor?

a. Nephron-sparing surgery
b. Local irradiation therapy
c. Three drug chemotherapy
d. Upfront nephrectomy

Answer to Question 3: d: Renal sparing surgical procedures are important in the management of bilateral Wilms tumor. The treatment of children with bilateral tumors must be individualized. The goals of therapy are to eradicate the entire tumor and at the same time preserve as much as normal renal tissue as possible with the hope of significantly decreasing the risk of chronic renal failure. Primary excision of the tumor should never be attempted in bilateral Wilms tumor.

## Further Reading

Alastair JWM, Cox S, Davidson A. Management of bilateral Wilms tumour. Pediatr Surg Int. 2017;33(7):737–45.

Shamberger RC. Renal tumors. In: Carachi R, Grosfeld L, editors. The surgery of childhood tumors. 3rd ed. Berlin: Springer; 2016. p. 189–218. Chapter 12.

Shamberger RC, Anderson JR, Breslow NE, Perlman EJ, et al. Long-term outcomes for infants with very low risk Wilms tumor treated with surgery alone in National Wilms Tumor Study-5. Ann Surg. 2010;251:555–8.

Shamberger RC, Guthrie KA, Ritchey ML, Haase GM, et al. Surgery-related factors and local recurrence of Wilms tumor in National Wilms Tumor Study 4. Ann Surg. 1999;220:292–7.

# Neuroblastoma

**Jagadeesh Ramdas**

**Key Points**

1) Biologically heterogeneous tumor with variable clinical presentation.
2) Most common solid intra-abdominal tumor in children.
3) Most common presentation is abdominal mass.
4) Can occur anywhere along the sympathetic chain from neck to pelvis.
5) Tumor markers are homovanillic acid (HVA) and vanillylmandelic acid (VMA).
6) Metastatic disease is associated with poor prognosis.
7) Treatment approach is risk stratified and includes surgical biopsy/resection, chemotherapy, radiation, and stem cell transplant in some cases.

Neuroblastoma arises from neural crest cells, most commonly in the adrenal medulla. It can occur at any site along the sympathetic nerve chain from neck to pelvis. It is the most common solid abdominal tumor in children. The tumor is part of the amine precursor and uptake decarboxylation (APUD) cell line, which produces catecholamines. Neuroblastoma is a member of the family of small round blue cell tumors of childhood. Over 90% of neuroblastoma produce elevated levels of catecholamines or catecholamine byproducts.

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1. Epidemiology
    a. Incidence: Second most common solid neoplasm in children after brain cancer. It accounts for 10% of all childhood cancers.
    b. Approximately 700 children are diagnosed each year. Six hundred fifty are neuroblastoma, others include ganglioneuroblastoma (more differentiated form) and pheochromocytoma. Fully differentiated form is benign ganglioneuroma.
    c. Most commonly occur in the adrenal gland (50%). Twenty-five percent (25%) occur in the retroperitoneal paraspinal ganglion, 20% in the posterior mediastinum, and 5% in the neck or pelvis.
    d. Most common cancer in infancy, incidence rate double that of leukemia in infants.
    e. Median age at diagnosis is <2 years old, for which 5-year survival is >83% as compared to 50–60% 5-year survival for older children. 35% of cases occur before one year of age.
    f. Seventy percent of children present with metastatic disease.

2. Pathophysiology: Originates from neural crest tissue. No known or established environmental influences or toxic exposures impact disease occurrences.
    a. Mostly sporadic, however, distinct familial types are also seen.
    b. Association of neuroblastoma with Hirschsprung disease and congenital central hypoventilation syndrome which are associated with PHOX2B gene mutations.
    c. Germline mutation in the ALK gene have been identified in familial neuroblastoma.

3. Clinical Features
    a. Presenting signs and symptoms of children with neuroblastoma are quite variable and reflect both the location of the primary tumor and the extent of disease.
    b. Children present with asymptomatic mass in the neck, thorax, abdomen, or pelvis.
    c. Horner's syndrome and superior vena cava syndrome are seen in cervical and apical thoracic tumors.
    d. Paraspinal tumors may present with spinal cord compression with neurological deficit.
    e. Metastatic disease typically involves bone and bone marrow, causing bone pain, and gait disturbance. Orbital metastasis causing proptosis and ecchymosis. Bilateral "racoon eyes" (periorbital ecchymosis) is associated with metastatic neuroblastoma.
    f. Patients with localized disease are often asymptomatic while patients with metastatic disease appear ill with systemic symptoms including fever, malaise, and anemia.

   g. Excess catecholamine secretion may result in hypertension, tachycardia, headache, and flushing episodes.
   h. Infants may present with skin deposits and profound hepatomegaly from metastatic deposits, which may result in respiratory distress and organ compromise.
   i. Rarely tumor secretion of vasoactive intestinal peptide (VIP secreting tumor) can result in profuse watery diarrhea.
   j. Opsoclonus/Myoclonus ataxia syndrome occurs coincident with neuroblastoma in about 4% of patients and believed to be immune mediated. It consists of random disconjugate eye movements, abnormal muscle jerks, and ataxia.

4. Diagnosis

   a. Pathological examination of tissue is necessary for the diagnosis.
   b. Bone marrow biopsy/aspirate may establish the diagnosis.
   c. Biopsy of the tumor may be necessary to establish diagnosis and obtain tissue for studies.
   d. Elevated urinary catecholamine excretion homovanillic acid (HVA) and vanillylmandelic acid VMA).
   e. Histological examination shows a small blue round cell tumor.
   f. Staging work up consists of CT of the chest, abdomen, and pelvis or MRI if spinal involvement suspected.
   g. Bony involvement evaluated with bone scan, metaiodobenzylguanidine (MIBG) scan or octreotide scan.
   h. Laboratory studies—Elevated serum ferritin, LDH, and neuron-specific enolase.

5. Prognostic Factors

   a. Staging of tumor: Stages I, II, and IV-S (infant <1-year-old with metastasis to skin, liver, and bone marrow) associated with favorable diagnosis. Stage III intermediate prognosis, and stage IV (metastatic disease) associated with poor prognosis.
   b. Age at presentation: Presentation at age <1 year associated with improved outcomes for all stages.
   c. Ratio of HVA to VMA >1 is associated with adverse prognosis.
   d. N-myc gene amplification (>10 copies) occurs in 20% of primary neuroblastic tumors and is strongly associated with the presence of metastatic disease and poor prognosis.
   e. Immunohistochemical staining of neuronal markers such as synaptophysin and neuron-specific enolase.
   f. Neuroblastic tumors include neuroblastoma, ganglioneuroblastoma, and ganglioneuroma. Ganglioneuroblastoma is better differentiated and associated with improved prognosis. Percentage of tumor which is ganglioneuroma relative to neuroblastoma component determines degree of differentiation.

g. Tumors are classified as favorable and unfavorable depending upon the degree of neuroblast differentiation, Schwannian stroma content, and mitosis-karyorrhexis index are part of staging.

6. Risk Group Classification

   The Children's Oncology Group neuroblastoma risk stratification is based on an international risk grouping system. Patients are assigned to low, intermediate, and high-risk categories based upon the analysis of age at diagnosis, INSS stage, histopathology, N-myc amplification status and DNA index.

## International Neuroblastoma Staging System

| Stage/ Prognostic group | Description |
|---|---|
| Stage 1 | Localized tumor with complete gross excision, with or without microscopic residual disease; representative ipsilateral lymph nodes negative for tumor microscopically (i.e., nodes attached to and removed with the primary tumor may be positive). |
| Stage 2A | Localized tumor with incomplete gross excision; representative ipsilateral nonadherent lymph nodes negative for tumor microscopically. |
| Stage 2B | Localized tumor with or without complete gross excision, with ipsilateral nonadherent lymph nodes positive for tumor. Enlarged contralateral lymph nodes must be negative microscopically. |
| Stage 3 | Unresectable unilateral tumor infiltrating across the midline, with or without regional lymph node involvement; or localized unilateral tumor with contralateral regional lymph node involvement; or midline tumor with bilateral extension by infiltration (unresectable) or by lymph node involvement. The midline is defined as the vertebral column. Tumors originating on one side and crossing the midline must infiltrate to or beyond the opposite side of the vertebral column. |
| Stage 4 | Any primary tumor with dissemination to distant lymph nodes, bone, bone marrow, liver, skin, and/or other organs, except as defined for stage 4S. |
| Stage 4S | Localized primary tumor, as defined for stage 1, 2A, or 2B, with dissemination limited to skin, liver, and/or bone marrow (by definition limited to infants younger than 12 months).[18] Marrow involvement should be minimal (i.e., <10% of total nucleated cells identified as malignant by bone biopsy or by bone marrow aspirate). More extensive bone marrow involvement would be considered stage 4 disease. The results of the MIBG scan, if performed, should be negative for disease in the bone marrow. |

A new staging system, the International Neuroblastoma Risk Group (INRG) staging system, based upon clinical criteria and tumor imaging has been developed and will facilitate a greater comparison of risk-based clinical trials conducted in different regions of the world and collaborative studies.

## International Neuroblastoma Risk Group Staging System

| INRG stage | Description |
|---|---|
| L1 | Localized tumors with no image-defined risk factors |
| L2 | Localized tumor with one or more image-defined risk factors |
| M | Distant Metastatic disease |
| MS | Metastatic disease in children below 18 months of age with metastasis limited to skin, liver and/or bone marrow (<10% involvement) |

1. Treatment
   A. Low-risk disease: Surgical excision alone is curative. Survival rates >95% for patients with stage 1 disease.
      a. High rate of spontaneous regression is seen in infants with stage 4S neuroblastoma. Newborns with small adrenal masses can resolve spontaneously with no interventions.
      b. Stage 2A/2B diseases have excellent survival outcomes with primary surgical excision alone as the treatment approach.
   B. Intermediate-risk disease- surgical excision along with tumor biology directed chemotherapy. Biologically favorable tumors require very minimal chemotherapy cycles, whereas for the patients with unfavorable biology will receive 4 or 8 cycles of chemotherapy based upon the status of the chromosomal alterations.
   C. High-risk disease: Survival for high-risk neuroblastoma has improved modestly in the last 20 years. Survival for those 1–5 years of age is about 20%.
      a. Treatment involves multimodality induction treatment with heavy intensity chemotherapy, radiation therapy, followed by intensification of consolidation therapy with autologous stem cell transplantation following myeloablative doses of chemotherapy.
      b. One of the challenges of high-risk neuroblastoma remains the microscopic residual refractory disease with multidrug-resistant microscopic disease.
         i. Targeted immunotherapy, which exploits tumor selectivity, has been explored. Disialoganglioside (GD2) is a potent immunotherapy target that is highly expressed in neuroblastoma tumors. AntiGD2 monoclonal antibodies using agents like human-murine chimeric antibody (CH14:18) has shown promising response with further improving the outcome of high-risk neuroblastoma.

        ii. Radio-iodinated meta-iodobenzyl guanidine (MIBG) is also being explored to provide targeted deliver of radiotherapy.
        iii. Tandem stem cell transplants have demonstrated moderate improvements.

D. Role of surgery

    a. Surgery is an integral part of neuroblastoma therapy. Diagnostic biopsies as well as gross total resection are critical for diagnosis, curative resection, and for obtaining sufficient tumor tissue. Sampling of nonadherent intracavitary lymph nodes is recommended for accurate staging.
    b. Complete surgical resection is for those with low-risk disease is considered curative in most cases.
    c. Surgical resection for intermediate-risk tumors may be unnecessary. Studies suggest that shrinkage of the tumor prior to surgery may reduce surgical risk.
    d. The overall surgical goal in high-risk patients with neuroblastoma is removal of as much of the tumor as possible after induction chemotherapy. Tumor resection should include as much preservation of full organ and neurologic function as possible.
    e. Neuroblastoma often encases major vessels. Resection of Stage 3 or Stage 4 neuroblastoma which involves major vessels requires judicious dissection in the subadventitial plane without sacrifice of major vessels. Injury to major vascular structures has been reported in up to 10%. Injury to involved organs occurs in about 5% of cases. The kidney is the most common affected organ with resultant nephrectomy.

## Additional Notes

Neuroblastoma is a heterogenous group of tumors. It is the most common solid intra-abdominal tumor of childhood and infancy. Tumors may undergo spontaneous regression or maturation to benign ganglioneuromas. Large-scale screening studies were carried out in Japan in the 1980s. Initial studies were promising, but subsequent studies determined that early detection did not result in improved outcomes since it detected many tumors which would resolve without treatment. Prognosis depends primarily on the age of presentation and the stage of the tumor. Metastatic neuroblastoma in children over the age of one is associated with a poor prognosis, although recent advances in stem cell transplant have been associated with marginally improved outcomes.

## Study Questions

Question 1: The most common solid intra-abdominal cancer seen in infants is

a. Acute Lymphoblastic leukemia
b. Acute myelogenous leukemia
c. Neuroblastoma
d. Medulloblastoma
e. Wilms tumor

Answer: (c.) Neuroblastoma. Neuroblastoma is the most common intra-abdominal cancer seen in infants and comprises 10% of all pediatric malignancies. Median age at diagnosis of neuroblastoma is 22 months of age with one third of patients diagnosed in the first year of life and 90% of children diagnosed before 5 years of age. Wilms tumor is the second most common in children.

Question 2: Which one of the neuroblastoma presentations is not considered an image-defined risk factor for surgical excision?

a. Tumor compressing the trachea
b. Tumor infiltrating the porta hepatis
c. Tumor crossing the midline
d. Tumor encasing the brachial plexus roots

Answer: (c.) Tumor crossing the midline. Image-defined risk factors (analogous to surgical-defined risk factors) are a set of predetermined risk factors which can increase the risk of surgical morbidity and limit the chances of complete gross surgical resection. Tumor crossing the midline equates with a Stage 3 neuroblastoma but is not considered an image-defined risk factor for surgical excision.

Question 3: All the following are accepted treatment modalities for high-risk neuroblastoma EXCEPT:

a. Stem cell transplantation
b. Radiation therapy
c. Radiofrequency ablation
d. Targeted Immunotherapy

Answer: (c.) Radiofrequency ablation. The outcome of high-risk neuroblastoma has steadily improved. Multimodality treatments, including chemotherapy, surgery, irradiation, tandem stem cell transplantation, immunotherapy, and differentiating therapy, are all used in the management of high-risk neuroblastoma. While intensification of consolidation therapy and the addition of post-consolidation immunotherapy both appear to improve Event Free Survival (EFS) for high-risk patients, a substantial number of children still progress during induction therapy to show evidence of persistent metastatic disease or relapse after completion of treatment.

Question 4: Which of the following are tumor markers for neuroblastoma (choose all that apply):

a. Metanephrine
b. Homovanillic acid
c. Alpha-feto protein
d. B HCG
e. Vanillylmandelic acid

Answer: (b.) and (e.): Homovanillic acid, and Vanillylmandelic acid. Neuroblastoma is a member of amine precursor and uptake family of tumors. Over 90% secrete catecholamines or byproducts. Metanephrine is a tumor marker used in pheochromocytoma but not for neuroblastoma. Homovanillic acid and vanillylmandelic acid are the tumor markers associated with neuroblastoma. Alpha-feto protein and B HCG are not associated with neuroblastoma.

## Further Reading

Brodeur GM. Spontaneous regression of neuroblastoma. Cell Tissue Res. 2018;372(2):277–86.
Cohn SL, Pearson AD, London WB, Monclair T, et al. The International Neuroblastoma Risk Group (INRG) classification system: an INRG Task Force report. J Clin Oncol. 2009;27(2):289–97.
Holcomb GW, et al. Neuroblastoma. In: Holcomb GW, Murphy JP, Ostlie DJ, editors. Ashcraft's pediatric surgery. 7th ed. Philadelphia, PA: Elsevier; 2020. p. 1010–30. Chapter 65.
Irwin MS, Naranjo A, Zhang FF, Cohn SL, et al. Revised neuroblastoma risk stratification system: a report from the Children's Oncology Group. J Clin Oncol. 2021;39(29):3229–41.
Kliegman RM, et al. Neuroblastoma. In: Kliegman RM, et al., editors. Nelson's pediatric surgery. 20th ed. Philadelphia, PA: Elsevier; 2017. p. 2461–4. Chapter 498.
Swift CM, Eklund MJ, Kraveka JM, Alazraki AL. Updates in diagnosis and treatment of neuroblastoma. Radiographics. 2018;38(2):566–80.
Ward E, DeSantis C, Robbins A, Kohler B, Jemal A. Childhood and adolescent cancer statistics, 2014. CA: Cancer J Clin. 2014;64(2):83–103.
Woods WG, Gao RN, Shuster JJ, et al. Screening of infants and mortality due to neuroblastoma. N Engl J Med. 2002;346:1041–6.

# Liver Tumors

**Jagadeesh Ramdas and Marc S. Lessin**

**Key Points**

1) Hepatoblastoma is the most common primary liver malignancy in children.
2) Elevated alpha-fetoprotein is commonly elevated in hepatoblastoma.
3) Treatment depends on accurate staging. Complete resection with clear margins is necessary. Extent of tumor involvement will dictate whether tumor resection or liver transplantation is appropriate.
4) Hepatocellular carcinoma is the second most common primary liver cancer in children, generally occurring in older children.
5) The most common benign tumors of the liver in children are hemangioma, hemangioendothelioma, mesenchymal hamartoma, adenoma, and focal nodular hyperplasia.

The majority of liver tumors in children are benign. Hepatoblastoma is the most common primary liver cancer in children. Curative treatment requires resection. Liver transplantation is an option for unresectable non-metastatic tumors. Neoadjuvant chemotherapy prior to definitive resection may facilitate resection of tumors that were otherwise non-resectable. Hepatocellular carcinoma occurs in

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older children and is generally associated with worse outcomes than hepatoblastoma. There are many benign tumors of the liver which often do not require treatment, but are a diagnostic dilemma to insure they are not malignant.

1. Epidemiology
    - A. Incidence: Malignant liver tumors in children account for 1.1% of all malignancies in children younger than 20 years of age with approximately 100–150 new cases annually in children in the United States. Liver tumors are broadly divided into:
        - i. Hepatoblastoma –predominant malignant tumor in young children.
        - ii. Hepatocellular carcinoma (HCC) is the predominant tumor type in adolescent children.
        - iii. Rare hepatic malignancies of childhood include undifferentiated embryonal sarcoma of the liver, angiosarcoma of the liver, and choriocarcinoma.
        - iv. Benign hepatic tumors include mesenchymal hamartoma, infantile hemangioendothelioma, hemangioma, focal nodular hyperplasia, adenoma, and angiolipoma
    - B. Etiology of hepatoblastoma
        - i. Associated congenital abnormalities include familial polyposis syndrome associated with APC gene mutation, Gardner syndrome, Beckwith–Wiedemann syndrome, and hemihypertrophy.
        - ii. Environmental factors-- Fetal alcohol syndrome, prematurity, prolonged parenteral nutrition, oral contraceptives, oral gonadotrophins, and parental exposure to known carcinogens
    - C. Etiology of hepatocellular carcinoma
        - i. Congenital abnormalities--tyrosinemia, biliary cirrhosis, glycogen storage disease, alpha 1 antitrypsin deficiency, hemochromatosis, and ataxia-telangiectasia
        - ii. Environmental factors- hepatitis B & C, viral infections, parasitic infections, aflatoxin, anabolic steroids, and environmental carcinogens
2. Pathophysiology
    - A. Epithelial type: Hepatoblastoma is classified as epithelial (56%) or mixed epithelial/mesenchymal (44%). Epithelial hepatoblastoma is single, massive, or multifocal, usually an extensively infiltrative, non-encapsulated tumor; gross features include fleshy homogenous tumors with hemorrhage and necrosis. The most common epithelial types are fetal (30%), embryonal (20%), and macrotrabecular in various combinations. Fetal histology is associated with the best prognosis. The most severe form is anaplastic.
    - B. Hepatocellular carcinoma tumors can vary from small tumors with focally infiltrative borders to massive and can also be multifocal or diffuse. Usually, they are more diffusely infiltrative.

3. Anatomy
   A. An imaginary line from the ductus venosus posteriorly (inferior vena cava) through the middle of the caudate lobe along the round ligament to the suprahepatic inferior vena cava divides the liver into right and left lobes.
   B. The liver is further divided into 8 segments (Couinaud segments) (Fig. 1)
   C. Blood supply:
      i. Arterial supply is from the portal vein and hepatic artery. Each of these vessels divides into right and left branches supplying respective lobes of the liver.
      ii. The portal vein provides 70–75% of the blood flow to the liver. The hepatic artery supplies the remaining 25–30%. The oxygenation sup-

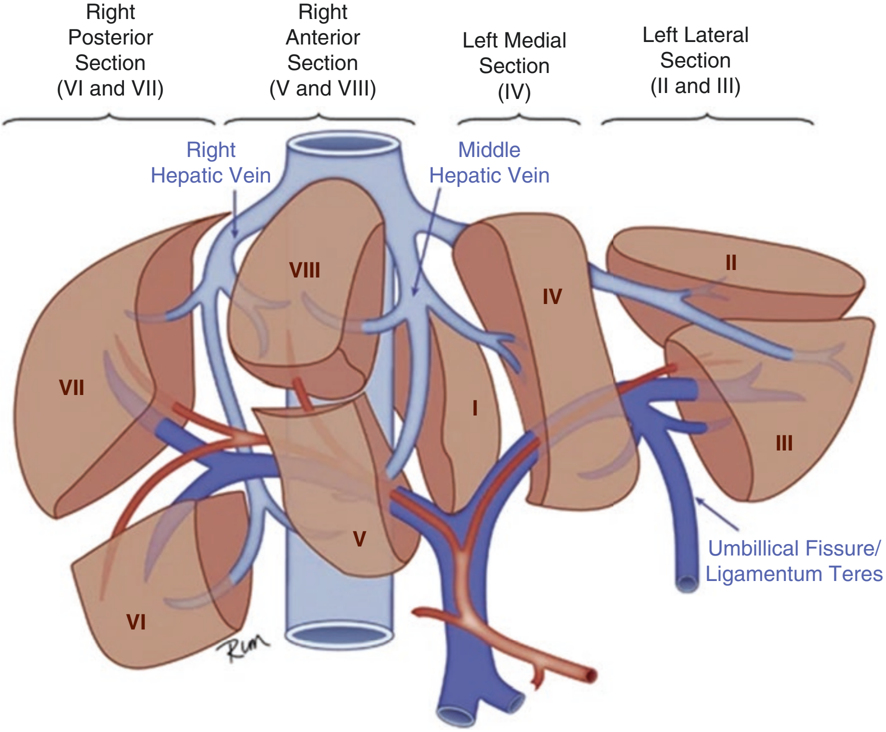
Right Posterior Section (VI and VII)
Right Anterior Section (V and VIII)
Left Medial Section (IV)
Left Lateral Section (II and III)
Right Hepatic Vein
Middle Hepatic Vein
VIII
IV
II
VII
I
III
V
VI
Umbillical Fissure/ Ligamentum Teres

**Fig. 1** Couinaud 8 segment anatomic division of the liver (Segments I–VIII) with comparison to the PRETEXT anatomic division of the liver into 4 Sections, as defined by the umbilical fissure and the right and middle hepatic veins. (Source: Lopez-Terrada D, Alaggio, de Davil MT, Czauderna P, Hiyama E, Katzenstein H, Leuschner I, Malogolowkin M, Meyers R, Ranganathan S, Tanaka Y, Tomlinson G, Fabre M, Zimmerman A, Finegold MJ. Towards an international pediatric liver tumor consensus classification: Proceedings of the Los Angeles COG liver tumors symposium. Modern Pathology 27: 472–491, 2014, Reprinted via Copyright Clearance Center: Springer Nature No. 5197390753328, 27 Nov 2021)

plied by each is approximately 50% as the hepatic artery has higher oxygen content.

- iii. The venous drainage is through the right, middle, and left hepatic veins which drain directly into the inferior vena cava.
- iv. The liver produces approximately one liter of bile per day in the adult. The right and left hepatic ducts coalesce into the common hepatic duct which then becomes the common bile duct after the cystic duct of the gallbladder enters. The drainage of the common bile duct is into the second part of the duodenum at the sphincter of Oddi.

4. Clinical features

   A. Most cases present with asymptomatic abdominal masses in the right upper quadrant. Occasionally associated with abdominal distension.
   B. Constitutional symptoms of anorexia, weight loss, fevers, and abdominal pain are less common but can be seen in hepatocellular carcinoma.
   C. Jaundice is very rare in pediatric liver tumors with the exception of rare cases of biliary tract tumors.
   D. Occasionally a patient may present with signs of precocious puberty secondary to abnormal secretion of human chorionic gonadotrophin.
   E. In patients with hepatocellular carcinoma associated with preexisting inflammatory or metabolic diseases, signs of hepatic cirrhosis are frequent and include hepato-splenomegaly as well as cutaneous telangiectasia.
   F. Hemoperitoneum secondary to tumor rupture can happen. Metastasis to the lung occurs in up to 20–30% of patients at the time of diagnosis for patients with hepatoblastoma and HCC.
   G. Peritoneal implants, extension into adjacent intra-abdominal structures and spread to distant lymph nodes, bones, and bone marrow can occasionally be seen.

5. Diagnostic Evaluation

   A. Laboratory Investigations

      i. Complete hematological evaluation- Thrombocytosis with platelet counts exceeding 500,000 per cu mm routinely observed in hepatoblastoma. Related to excess production of C-megakaryocyte proliferating ligand (C-MPL) from the hepatoblastoma tumor tissues.
      ii. Liver enzymes and alkaline phosphatase may be elevated in some patients with HCC but are frequently normal in hepatoblastoma.
      iii. Serum alpha-fetoprotein (AFP) is the most sensitive tumor marker for diagnosis and clinical evaluation. AFP levels are almost always elevated in children with hepatoblastoma and elevated in 50–60% of children with hepatocellular carcinoma. AFP is an excellent biological marker to determine tumor activity.
      iv. Beta-HCG is abnormally elevated in those cases presenting with isosexual pseudo-precocious puberty.

    v. Fibrolamellar variant of HCC can be associated with an abnormality of Vitamin B12 binding protein, which can be used to monitor disease status and response to therapy. Some studies show improved outcomes with fibrolamellar variant.

6. Radiographic Studies
   A. US imaging—reliable and noninvasive imaging technique in establishing the presence of an intrahepatic mass.
      i. Contrast-enhanced ultrasound (CEUS) has been used to determine benign versus malignant liver tumors. Gas-containing contrast agents are injected and visualized in real time.
   B. CT scan of the abdomen and chest—most commonly used imaging study to determine both local and distal extent of tumor involvement.
   C. MRI can also be used.

7. Tumor staging--based on operative findings

   Stage I—Tumors that are completely resected and have a typical fetal histology
   Stage II—Tumors are grossly resected with evidence of microscopic residual disease
   Stage III—(Unresectable)- considered as unresectable tumors upfront by the surgeon. Also includes partially resected tumors with measurable tumors left behind and lymph node involvement
   Stage IV—Evidence of measurable residual disease to lungs or other organs

8. Treatment
   a. Surgery: Complete tumor resection is the cornerstone of therapy for liver tumors and offers the only realistic chance of long-term disease-free survival.
   b. Initial Measures: Biopsy- combination of clinical findings along with elevated alpha-fetoprotein levels (AFP) strongly suggest the clinical diagnosis of hepatoblastoma; however, pathological determinants of the diagnosis remain the crucial step.
   c. Subsequent Measures:
      i. Tumor resection- irrespective of whether definitive resection surgery is done at initial diagnosis or upon subsequent second surgery following chemotherapy, complete resection of tumor represents the main goal of any treatment strategy. Complete tumor resection is possible at diagnosis in 30–40% of cases, while it becomes possible at different time points after chemotherapy in 70–80% of cases overall.

ii. In cases of tumors located in close proximity to main liver vessels like portal vein and hepatic veins, and where resection would require peeling tumors of the vessels, microscopic residual of the tumors is inevitable. Prognostic significance of leaving microscopic residual tumor is not clearly established. Microscopic residual tumor does not necessarily carry an adverse prognosis.

  iii. Surgical resection is also advocated for those patients with isolated lung metastases remaining after chemotherapy.
  iv. Hepatic surgery for tumor removal always implies a small but significant risk of uncontrolled blood loss. Hence, tumor resection surgery should only be undertaken in centers, which have skilled and well-experienced surgeons as well as anesthesiology teams. Standard hepatic resection techniques should be recommended including segmentectomies, hemi-hepatectomies, and extended hemi-hepatectomies in advanced cases. Gross total resection offers realistic hope of cure for both hepatoblastoma and hepatocellular carcinoma.

*Chemotherapy*

A. Chemotherapy plays an important role and significant role in the improvement of survival of children with hepatoblastoma. Agents with therapeutic efficacy include cisplatin, vincristine, 5-fluorouracil, and doxorubicin. Other agents including irinotecan, topotecan, etoposide have also been found to be useful in recurrent settings.
B. Chemotherapy is given either as adjuvant chemotherapy following initial surgical resection if feasible for low or intermediate risk hepatoblastoma, and has helped to drastically reduce the tumor recurrence rate.
C. Neoadjuvant chemotherapy with the objective of increasing the number of patients for whom complete surgical resection is feasible and reducing surgical morbidity provides more time for making definitive surgical plans, and has been now more widely adopted based upon the SIOPEL (liver tumor study group of the International Society of Oncology) studies which established the PRETEXT staging system to determine those who should undergo preoperative chemotherapy.
D. The PRETEXT (PRE-Treatment EXTent of disease) stage refers to the number of sectors involved (i.e., 1 through 4). The liver is divided into four sectors consisting of the lateral left lobe (Couinaud's segments 2 and 3), medial left lobe (segment 4), anterior right lobe (segments 5 and 8) and posterior right lobe (segments 6 and 7) (Table 1).
E. When the tumor is limited to one or two sectors, partial hepatectomy is appropriate at the outset. However, when the tumor is more extensive, preoperative chemotherapy with a cisplatin, 5-fluorouracil and vincristine-based regimen is recommended in an effort to shrink the tumor sufficiently for complete resection. If the tumor has not sufficiently regressed after two cycles to permit resection, it is not warranted to continue intensification with further chemotherapy, but rather list for liver transplantation.
F. There is minimal role of chemotherapy in hepatocellular carcinoma (HCC).

  i. Chemoembolization and radiofrequency ablation are both used in the treatment of HCC.

**Table 1** PRETEXT classification of hepatoblastoma (Source: Adapted from Meyers R, Tiao G, de Ville de Goyet J, Superina R, Aronson DC. Hepatoblastoma state of the art: Pre-treatment extent of disease, surgical resection guidelines and the role of liver transplantation. Hunger SP (ed.) Hematology and Oncology, Current opinion in Pediatrics, 26(1): 29–36, 2014)

| **Stage** | **Hepatoblastoma extent** |
|---|---|
| PRETEXT I | One section involved |
| PRETEXT II | One or two sections involved; two adjoining sections are tumor free |
| PRETEXT III | Two or three sections involved; one adjoining section is tumor free |
| PRETEXT IV | All sections involved |
| **Annotation** | **Meaning** |
| V | Venous involvement of the retrohepatic vena cava or all hepatic veins |
| P | Portal vein involvement |
| E | Extrahepatic involvement of adjacent organs |
| M | Distant metastatic disease |
| C | Involvement of caudate lobe |
| F | Multifocal tumor nodules |
| R | Tumor rupture at diagnosis |

   ii. Sorafenib a multikinase inhibitor with angiogenic, pro-apoptotic, and raf kinase inhibitory activity has shown to improve overall survival for patients with advanced HCC.
   iii. The role of radiation therapy in the treatment of hepatoblastoma and HCC has not been established.

G. Liver transplantation: Liver transplantation has added a new dimension to the treatment of hepatoblastoma and represents a feasible alternative for the treatment of patients with unresectable tumors.

   i. Tumors are deemed primarily unresectable at presentation if there is multifocal involvement with limited hepatic reserves after partial hepatectomy, or may be considered unresectable because of limited initial response to chemotherapy, or presence of extensive tumor infiltration, or thrombus in the hepatic or portal veins.
   ii. Early referral to a liver transplant center is recommended for tumors such as central PRETEXT III with close proximity of the tumor to the main hepatic veins and/or main portal branches or PRETEXT IV.
   iii. Options for transplantation include the traditional deceased donors or more recently living donor transplants.
   iv. Close communication is essential between oncology team, surgeon, and liver transplant team as definitive surgical resections or transplants are done after 2 or 3 cycles of initial chemotherapy to determine whether resection or transplantation is needed.

   v. Close evaluation by imaging for any perceived metastatic sites at both intraabdominal as well as distant metastatic sites should be done initially as well as after few cycles to determine transplant eligibility.

H. New methods of local tumor control in the form of hepatic artery chemoembolization, radiofrequency ablation, Y-90 microsphere radio embolization to target tumor thrombus are all being explored and showing promising responses, and improving the chance of complete resection.

## Benign Liver Tumors

A. Hemangioma and hemangioendothelioma
   i. Most common benign hepatic tumor of childhood.
   ii. Presents shortly after birth.
   iii. Usually asymptomatic but large lesions can result in hepatomegaly, thrombocytopenia, congestive heart failure, respiratory distress, and gastric outlet obstruction.
   iv. Treatment for asymptomatic lesions is observation. Symptomatic lesions with coagulopathy, respiratory distress, or vomiting are treated with propranolol and/or steroids. Embolization may be necessary for large shunts; although, this poses risks of embolization.

B. Mesenchymal hamartoma
   i. Second most common benign tumor in childhood
   ii. Typically presents before the age of 2
   iii. Strong preponderance for right lobe of liver
   iv. May be solid, cystic, or mixed
   v. Treatment is complete resection. May undergo malignant transformation or harbor co-existing embryonal sarcoma.

C. Focal nodular hyperplasia
   i. Often discovered incidentally. May present with abdominal pain, mass, weight loss, or early satiety.
   ii. Lesions < 5 cm that are asymptomatic with classic findings on CT can be watched. Lesions may regress over time.
   iii. Percutaneous biopsy may be helpful but is only diagnostic in about one-half.
   iv. Symptomatic lesions, those < 5 cm or showing interval growth should undergo resection.

D. Adenoma
   i. Resemble focal nodular hyperplasia on imaging
   ii. Most common in young women. Associated with birth control pills or anabolic steroids. Cessation of these medications may result in regression.

iii. Usually solitary lesion
iv. Adenomas > 5 cm are at risk for rupture with bleeding
v. Malignant transformation is rare. Male sex, size of the tumor and certain subtypes are at risk.
vi. Treatment for large adenomas without regression is resection. Radiofrequency ablation may also be successful.

## Additional Notes

Liver tumors in children are comprised of a variety of malignant and benign diseases. Hepatoblastoma is the most common primary malignancy in childhood. The mainstay of treatment requires resection. Survival has increased with the application of preoperative chemotherapy following biopsy. Liver resections are a major operative undertaking and should be performed at centers with experience in hepatobiliary surgery. New and evolving energy devices such as Ligasure, Harmonic Scalpel, CUSA, and Aquamantys should be used and be available. Liver transplant has become an important part of treatment for unresectable tumors. Intraoperative ultrasound may be helpful in delineating the extent of tumor to attain clear margins. Benign liver tumors create a diagnostic dilemma as there may be difficulty in distinguishing from malignant tumors. New imaging technologies have improved diagnosis, but biopsy may be necessary in some cases.

## Study Questions

1) All the following are known risk factors associated with pediatric liver tumors except:

   a. Familial Polyposis
   b. Beckwith–Wiedemann Syndrome
   c. Hemi hypertrophy
   d. Trisomy 21 –Down syndrome

Answer to Question 1: (d.) Trisomy 21 -Down syndrome. Children with Beckwith–Wiedemann syndrome (BWS) and hemihypertrophy (HH) are at higher risk of embryonal tumor development such as Wilms tumors, embryonal rhabdomyosarcoma as well as hepatoblastomas. The association of familial adenomatous polyposis (FAP) and hepatoblastomas is related to the alteration in the degradation of B-catenin gene, which is regulated by the APC gene, which drives the production of polyposis.

Question 2: What Staging system is used as a guide to decide about surgical resectability of liver tumors

a. UNOS staging
b. MILAN Staging
c. PRETEXT Staging
d. COG Staging

Answer to Question 2: (c.) PRETEXT Staging. Pre-treatment evaluation of Tumor Extension: (PRETEXT) relies on radiologic staging; using the main hepatic veins and portal veins to identify the number of liver sectors involved by tumor: In the PRETEXT system Liver is divided into 4 sectors-an anterior and posterior sector on the right lobe and a medial and lateral sector on the left lobe. Tumors are considered as Pre Text1- when 3 adjoining sectors are free and only 1 is involved by the tumors; PRETEXT II- when 2 adjoining sectors are free and 2 are involved, Pre Text III- when just 1 sector is free and 3 involved; PRETEXT IV- when there are no free tumor sectors.

Question 3: All the following modalities of treatments are used in Hepatoblastoma except:

a. Neoadjuvant chemotherapy
b. Surgical Resection
c. Radiation therapy
d. Liver Transplantation

Answer to Question 3: (c.) Radiation therapy. The role of radiation therapy in the treatment of hepatoblastoma as well as hepatocellular carcinomas has not been established. Neoadjuvant chemotherapy with the aim of increasing the number of patients who can achieve a complete surgical resection is a well-accepted standard of care, helps reduce surgical morbidity, and provides more time for making definitive surgical plans including liver transplantation when indicated.

Question 4: Which of the following is the most common benign liver tumor in childhood?

a. Adenoma
b. Focal nodular hyperplasia
c. Hemangioma
d. Mesenchymal hamartoma

Answer: (c.) Hemangioma. Hemangioma is the most common benign tumor in childhood. Most hemangiomas will regress over time, but treatment may be required for coagulopathy, high output shunting with congestive heart failure, protracted vomiting, or respiratory compromise.

## Further Readings

Czauderna P, Lopez-Terrada D, Hiyama E, et al. Hepatoblastoma state of the art: pathology, genetics, risk stratification, and chemotherapy. Curr Opin Pediatr. 2014;26(1):19–28.

Czauderna P, Otte JB, Aronson DC, et al. Guidelines for surgical treatment of hepatoblastoma in the modern era--recommendations from the Childhood Liver Tumour Strategy Group of the International Society of Paediatric Oncology (SIOPEL). Eur J Cancer. 2005;41(7):1031–6.

Dietrich CF, Nolsoe CP, Barr RG, Berzigotti A, et al. Guidelines and good clinical practice recommendations for Contrast-Enhanced Ultrasound (CEUS) in the Liver-Update 2020 WFUMB in Cooperation with EFSUMB, AFSUMB, AIUM, and FLAUS. Ultrasound Med Biol. 2020;36(10):2579–604.

Grosfeld JL, Otte JB. Liver tumors in children. In: Carachi R, Grosfeld JL, editors. The surgery of childhood tumors. 3rd ed. Berlin: Springer; 2016, Chapter 16. p. 269–302.

Meyers RL, Tiao G, de Ville Goyet J, et al. Hepatoblastoma state of the art: pre-treatment extent of disease, surgical resection guidelines and the role of liver transplantation. Curr Opin Pediatr. 2014;26(1):29–36.

# Leukemia

**Michal A. Miller**

**Key Points**

1) Acute leukemias are the most common pediatric malignancy. They are subdivided into acute lymphoblastic leukemia (ALL), which is more common, and acute myeloblastic leukemia (AML).
2) Patients with newly diagnosed leukemia can suffer the life-threatening condition of tumor lysis syndrome, in which multiple lysing tumor cells result in hyperkalemia, hyperphosphatemia, elevated uric acid, and renal failure.
3) As leukemia is treated with chemotherapy, the most common surgical need of a child with leukemia is tunneled in-dwelling central line placement for the administration of chemotherapy. The surgeon must address the anemia, neutropenia, and thrombocytopenia suffered by children with leukemia to avoid the surgical complications of cardiovascular collapse, infection, and hemorrhage.

1. Epidemiology
    a. Acute leukemias are the most common pediatric malignancy.
    b. Acute lymphoblastic leukemia (ALL) is the most common leukemia (75%) followed by acute myeloblastic leukemia (AML).
    c. Annually, approximately 3500 new cases of leukemia are diagnosed in the USA in children < 20 years of age.

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   d. Peak incidence is 2 to 3 years of age for ALL followed by a decreasing incidence until 10 years of age when the incidence climbs with increasing age.
   e. There is an increased incidence in children with congenital syndromes. The most common are Downs, Neurofibromatosis, Shwachman syndrome, Bloom syndrome, ataxia telangiectasia.

2. Clinical Presentations:
   a. Children present with constitutional symptoms of fevers, weight loss, night sweats, chills and a mixture of signs and symptoms related the bone marrow failure and tumor burden. Thrombocytopenia presents with petechiae, mucosal bleeding, easy bruising, and less commonly as a life-threatening hemorrhage. Anemia can lead to pallor, hypersomnia, fatigue, irritability, and poor feeding in infants, tachycardia with varying degrees of cardiovascular compromise. Life-threatening infections related to the neutropenia.
   b. The leukemia cell infiltration results in adenopathy, hepatosplenomegaly, and bone pain.
   c. Urgent complications encountered at diagnosis related to the tumor burden need to be anticipated.
      i. Tumor lysis syndrome: Spontaneous or therapeutic lysis of tumor cells leads to the rapid release of intracellular cytokines, potassium, and phosphorus. Rapid cell turnover results in the buildup of uric acid, which can precipitate in the renal tubules resulting in renal failure. Treatment is anticipatory. Administer hyperhydration to maintain urine output of > 2ml/kg/hr. This can usually be achieved with 5% dextrose one half normal saline or normal saline given two times the patient's intravenous fluid maintenance rate. No supplemental potassium is given. Monitor input and output for the development of oliguric/anuric acute renal failure.
         1. Allopurinol to reduce uric acid production.
         2. Aggressive treatment of electrolyte disturbances: hyperkalemia, and hyperphosphatemia and hypocalcemia.
         3. Fever and neutropenia. Patients with neutropenia absolute neutrophil count (ANC) < 1000 need urgent antibiotic coverage with empiric broad-spectrum antibiotics. Cefepime is the front-line therapy.
         4. Symptomatic or prophylactic platelet transfusions are given for thrombocytopenia.
      ii. Leukostasis: Most commonly encountered in AML patients with a WBC of > 100,000 and ALL patients with >300,000. Can present with respiratory insufficiency and/or stroke-like signs. Leukostasis requires urgent initiation of chemotherapy and/or leukapheresis.
      iii. Mediastinal mass and airway compromise is most common in T cell leukemia or lymphoma.
      iv. Spinal cord compression from tumor mass effect.

- d. Urgent pediatric oncology consult should be obtained on any patient suspected of having leukemia or lymphoma. These patients frequently decline rapidly.

3. Diagnosis:
    - a. The diagnosis is usually suspected or evident when a CBC with differential shows blast cells and/or varying degrees of pancytopenia.
    - b. Bone marrow aspiration is needed to confirm and subtype the leukemia.
    - c. Cerebrospinal fluid (CSF) cytology is obtained to determine the extent of the disease.
    - d. Labs most helpful at diagnosis are BMP, LFT, CBC with differential, uric acid, phosphorus, LDH, PT, PTT, fibrinogen and a type and screen.

4. Treatment:
    - a. The prognosis and treatment are dependent on the classification and the risk stratification of the leukemia subtype.
    - b. Children with leukemia will usually require a tunneled intravenous line for chemotherapy, such as a Broviac (Becton, Dickenson, and Company, Franklin Lakes, NJ), or with an attached subcutaneous reservoir such as a Port-a-Cath (Smiths Medical, Dublin, OH).
        - i. A single lumen catheter is used for patients with ALL.
        - ii. A double lumen catheter is used for patients with AML.
        - iii. This decision should be discussed with the treating oncologist, who may also do a bone marrow biopsy at the time of catheter placement under general anesthesia.
        - iv. Parents of children receiving an intravenous line should be warned not to give their child non-steroidal anti-inflammatory medications such as ibuprofen for analgesia, because these medications will carry a risk of stress ulcer when chemotherapy causes thrombocytopenia.
    - c. The Children's Oncology Group (COG) has developed a complex stratification for ALL and AML.
        - i. Subtype by flow cytology. Determines B cell ALL, T cell ALL and Myeloid phenotypes.
        - ii. The initial risk stratification for ALL starts with the National Cancer Institute criteria age > 1-year-old and < 10-years-old with a white blood cell count on presentation of < 50,000/μL are considered low risk. Age outside of that range and/or a WBC > 50,000 is high risk.
        - iii. Cytogenetics. For ALL, unfavorable cytogenetics include hypodiploidy (< 45 chromosomes), Philadelphia chromosome (BCR-ABL1), and the translocations t (9;22), t (1;19), and t (4;11). Favorable cytogenetics for ALL include combined trisomies of chromosome 4 and 10 and ETV6/RUNX1 fusion. For AML, unfavorable cytogenetics include

Monosomy 5 or 7 FLT3, Favorable cytogenetics for AML include: inversion of chromosome 16, and t (8;21), t (15;17), CEBPA and NPM1 mutations.

   iv. Central nervous system (CNS) and/or testicular involvement.
   v. Response to the therapy.

d. Chemotherapy for ALL consists of phases- induction, consolidation, interim maintenance, delayed intensification, and a prolonged maintenance. The first 4 phases consist of intensive chemotherapy that results in severe immunosuppression and pancytopenia. The maintenance phase last 2 to 3 years and is fairly mild. The patient's blood counts are generally fairly good, and the child can undergo minor procedures safely. Therapy for AML is much more intense and consists of 6 cycles of chemotherapy with prolonged hospitalizations for severe neutropenia. In the presence of unfavorable features and the availability of a marrow donor, many children will undergo bone marrow transplantation. The prognosis for children with ALL is good. The long-term survival rate for children with ALL, as a group, is 70–80%. The cure rate for the low-risk subset of ALL with favorable features is a > 90%. The prognosis for children with AML lower than it is for ALL.

## Additional Notes

Although the curative treatment for leukemia is non-operative, children with leukemia will often need the care of a surgeon for placement of an indwelling tunneled intravenous catheter to receive chemotherapy. The surgeon will likely see the child at the end of chemotherapy for another operation to remove the catheter. At anytime along the course of treatment, a catheter complication or failure may result in the need for revision of the catheter.

The course of chemotherapy will vary based on the type of leukemia being treated, the risk stratification, and the response to therapy. Chemotherapy for ALL consists of phases- induction, consolidation, interim maintenance, delayed intensification, and a prolonged maintenance. The first 4 phases consist of intensive chemotherapy that results in severe immunosuppression and pancytopenia. The maintenance phase lasts 2 to 3 years and is fairly mild. The patient's blood counts are generally fairly good, and the child can undergo minor procedures safely. Therapy for AML is much more intense and consists of 6 cycles of chemotherapy with prolonged hospitalizations for severe neutropenia. In the presence of unfavorable features and the availability of a marrow donor, many children will undergo bone marrow transplantation. The prognosis for children with ALL is good. The long-term survival rate for children with ALL, as a group, is 70–80%. The cure rate for the low-risk subset of ALL with favorable features is > 90%. The prognosis for children with AML lower than it is for ALL.

## Study Questions

1) Question 1: Which is not a common presenting symptom of Leukemia?

   a. Fever
   b. Weight loss
   c. Cervical mass
   d. Night sweats
   e. Easy bruising

Answer: (c.) Cervical mass. Leukemia is the most common pediatric malignancy. It is a tumor of the bone marrow, and the hematopoietic system specifically. The "tumor" is uncontrolled, inappropriate growth of abnormal white blood cells. This growth results in the reduced production and circulating numbers of normal cells produced in the bone marrow. Anemia from reduced red cell mass results in fatigue, pallor, hypersomnia, irritability, poor feeding, tachypnea, and cardiovascular compromise. Neutropenia results in susceptibility to life-threatening infections. Thrombocytopenia results in hypercoagulability, easy bruising, and bleeding. Neck mass is not a typical feature of Leukemia.

2) Question 2: Which is not an indicated therapy for a child presenting with tumor lysis syndrome?

   a. Potassium administration
   b. Hyperhydration
   c. Allopurinol
   d. Antibiotic coverage
   e. Correction of electrolyte disturbances

Answer: (a.) Potassium administration. Patients with newly diagnosed leukemia can present with the life-threatening emergency of tumor lysis syndrome. This can occur spontaneously as part of disease progression or can be precipitated by initial cytotoxic chemotherapy. Lysis of tumor cells results in massive release of cytokines, potassium, and phosphorus, Rapid cell turnover results in buildup of uric acid, which can precipitate in the renal tubules resulting in renal failure. Patients are treated with hyperhydration to assist with renal filtration. Electrolyte disturbances are corrected, but no potassium is given because tumor lysis syndrome characteristically causes hyperkalemia. Allopurinol is given to reduce uric acid production. Additionally, patients will need prophylactic empiric broad-spectrum antibiotics to protect them from the vulnerability to infection caused by neutropenia. Patients will also need treatment with platelet transfusions to treat thrombocytopenia and avoid bleeding.

3) Question 3: Which features in a patient with Acute lymphoblastic leukemia (ALL) indicate high risk disease?
   a. Presenting WBC < 50,000/μL
   b. Age between 1-year-old and 10 years old
   c. Combined trisomies of chromosome 4 and 10
   d. Hypodiploidy (< 45 chromosomes)

Answer: (d.) Hypodiploidy (< 45 chromosomes) Children's Oncology Group (COG) has developed a risk stratification for ALL. Age between 1-year-old and 10 years old is low risk; age outside that range is high risk. Initial CBC with WBC < 50,000/μL is low risk; WBC > 50,000/μL is high risk. Cytogenetic features that are low risk are combined trisomies of chromosome 4 and 10, and ETV6/RUNX1 Fusion. Cytogenetic features that are high risk are hypodiploidy (< 45 chromosomes, Philadelphia chromosome (BCR-ABL1), and the translocations t (9;22), t (1;19), and t (4;11). Involvement of the central nervous system (CNS) and/or testicles is high risk. Poor response to therapy is high risk.

## Further Reading

Children's Oncology Group (2021) Children's oncology group clincial protocol summaries. https://www.childrensoncologygroup.org/family-protocol-summaries. Accessed 18 Oct 2021

Lanzkowsky P, Lipton JM, Fish JD, editors. Lanzkowsky's manual of pediatric hematology and oncology. London: Academic Press; 2016.

Orkin SH, Nathan DG, Ginsburg D, Look AT, Fisher DE, Lux S. Nathan and Oski's Hematology and oncology of infancy and childhood. Philadelphia, PA: Elsevier; 2015.

Pizzo PA, Poplack DG. Principles and practice of pediatric oncology. Philadelphia, PA: Wolters Kluwer; 2016.

# Rhabdomyosarcoma

**Jagadeesh Ramdas**

**Key Points**

1) Rhabdomyosarcoma is a neoplasm of skeletal muscle origin with major histologic subtypes of embryonal and alveolar.
2) Rhabdomyosarcoma can arise in diffuse locations throughout the body including head and neck, genitourinary, trunk, extremities, and retroperitoneum.
3) Multimodality treatment approach with surgical resection, radiation therapy, and chemotherapy is necessary to achieve cure.

Soft tissue sarcomas are a heterogeneous group of malignant connective tissue tumors that arise in soft tissue sites throughout the body. In children, the most common soft tissue sarcoma is rhabdomyosarcoma (RMS). All the other sarcomas are individually rare and often grouped together as Nonrhabdomyosarcoma soft tissue sarcoma (NRSTS).

1. Epidemiology
    a. Incidence: Soft tissue sarcoma comprises 7% of all childhood cancer cases. RMS is 40% of soft tissue sarcomas in children and occurence is 4.5 cases per million children [1]. RMS accounts for 3% of all childhood cancers. RMS is most common in younger children with approximately 350 new cases

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reported in the United States each year. Incidence rates are slightly higher in boys than in girls for all ages.

- b. Predisposing risk factors: Most of soft tissue sarcoma develop in children without any recognized risk factors. Risk of rhabdomyosarcoma is higher with certain syndromes: Neurofibromatosis type 1, Beckwith–Wiedemann syndrome, Li–Fraumeni syndrome, Cardio-facio-cutaneous syndrome, and Costello syndrome.
- c. Non-Rhabdomyosarcoma soft tissue sarcoma- specific type malignant peripheral nerve sheath tumors may arise in benign neurofibromas in patients with NF-1.
- d. Children with hereditary Retinoblastoma who harbor germline Rb mutations are at increased risk of development of sarcomas.

2. Pathophysiology

   - a. The hallmark of rhabdomyosarcoma is evidence of skeletal muscle differentiation. Muscle-related immunohistochemical stains (myogenin, MyoD, muscle-specific actin, myoglobin, and/or desmin) are needed to differentiate from other small, round blue cell neoplasms that occur in children and adolescents. In addition, molecular analysis for the detection of translocations and other gene expression markers is becoming increasingly important.
   - b. Subtypes of Rhabdomyosarcoma [2, 3]

     - i. Embryonal Rhabdomyosarcoma (ERMS) is the most common histologic variant (approximately 36–90%). It is characterized by primitive spindle cells often with myxoid areas. ERMS occurs in younger children, commonly in the head/neck region, orbit, and genitourinary system.

       1. The botryoid variant of embryonal RMS (6–8%) typically arises in hollow organs, like the vagina or bladder, and presents as a grapelike, protruding mass. It carries a good prognosis.
       2. Spindle cell and sclerosing RMS (7%) is a rare variant of ERMS (leiomyomatous with cross striations)

     - ii. Alveolar Rhabdomyosarcoma (ARMS) is less common (approximately 20–40%). ARMS is composed of round or oval cells that form alveolar spaces surrounded by a framework of fibrous septae. Usually confers an unfavorable prognosis. In ARMS there is a typical chromosomal translocation t(2:13)(q35;q14) and a variant, t(1:13)(p36;q14), which fuses either the PAX3 gene or the PAX7 gene with the C-terminal transactivation domain of FOXO1 [2]. ARMS tends to occur in adolescents and in the extremities.
     - iii. Rare histologic subtypes of RMS are undifferentiated (3%), pleomorphic (1%), and other (5%).

3. Clinical features

   - a. Rhabdomyosarcoma has a heterogeneous presentation. Can occur anywhere and is not associated with any organ or body site and not limited to muscle.

Presents as painless mass. However, other symptoms can occur depending upon the location and compression or invasion of adjacent structures. For example, orbital tumors cause proptosis, nasopharyngeal and paranasal tumors cause airway obstruction, sinusitis, pain, epistaxis, and cranial nerve palsies. Bladder and prostate tumors present with hematuria, urinary obstruction, and constipation.

b. Most common primary sites of RMS are: [3]

    i. Genitourinary system including urinary bladder, prostate, vagina, and uterus (24%)
    ii. Extremities upper and lower (19%)
    iii. Parameningeal (16%)
    iv. Retroperitoneum/Pelvis (12% cases)
    v. Head and neck (10%)
    vi. Orbit (9%)
    vii. Trunk (8% cases)
    viii. Other (22%)

c. At diagnosis, 85% of cases are localized to the primary site with no evidence of distant spread. Fifteen percent of cases present with metastatic disease, primarily into lungs, bone marrow, bone, and lymph nodes.

d. Patterns of spread:

    i. Lymphatic: 40% of paratesticular and 20% of extremity tumors.
    ii. Hematogenous: 10–20% at diagnosis (lung, bone, bone marrow, liver).
    iii. CNS extension: 50% of parameningeal (cranial nerve palsies, erosion of cranial bone, direct intracranial growth).

5. Diagnostic Evaluation

    a. Complete physical exam with a precise definition of tumor site and size and a careful assessment of regional lymph nodes
    b. Laboratory studies including complete blood count, liver function testing, renal function testing, electrolytes, and coagulation parameters.
    c. Imaging: Magnetic Resonance Imaging (MRI) or Computed Tomography (CT) scan of the primary lesion, chest X-ray, and chest CT scan.
    d. Bone marrow aspiration and biopsy.
    e. PET scan is also recommended.
    f. Open surgical biopsy is preferred method to diagnose RMS in order to provide sufficient material for histopathological interpretation and for additional cytogenetic and molecular biology studies.
    g. Retroperitoneal lymph node sampling is necessary for paratesticular high-risk patients (>10-years-old, primary tumor size >5cm).

6. Classification and Staging

    a. Extent of disease is one of the most important predictors for outcome in children with RMS. Intergroup Rhabdomyosarcoma Study Group ((IRSG) cooperative group have designated a clinical grouping system, which is used to assess the extent of the tumor. Clinical group is determined by the surgical resectabilty of the primary tumor and presence of metastatic disease. From the pathological perspective, the principal determinant of poor prognosis is the presence of alveolar histologic subtype.

b. Patients with completely excised tumors (Group I), those with tumors arising in favorable sites (Stage 1), and those without regional node involvement (N0) consistently have the most favorable outcomes. Patients with metastatic disease (Group IV, stage 4) including those with alveolar histology, and older than 10 years with embryonal RMS have inferior outcomes.

c. IRS Grouping System: [4]

   i. Group I: Localized disease completely resected.
   ii. Group II: Gross total resection with evidence of regional spread.
      1. (A) Gross resection of tumor with microscopic residual disease.
      2. (B) Complete resection of primary tumor, regional nodes involved.
      3. (C) Regional disease with involved nodes: microscopic residual disease and/or histologic involvement of the dissected regional node most distal to the primary tumor site.
   iii. Group III: Incomplete resection with gross residual disease (biopsy only)
   iv. Group IV: Distant metastatic disease (lung, liver, bone, bone marrow, brain, and distant muscle and nodes)

d. IRS-Modified TNM Stage

   i. Stage 1: Describes a local tumor in the orbit; head and neck area, except for parameningeal sites; or a genitourinary tract tumor, except for in the bladder or prostate.
   ii. Stage 2: Describes a small local tumor in any part of the body not in Stage 1. The tumor is smaller than 5 cm, and there is no spread to regional lymph nodes.
   iii. Stage 3: Describes a local tumor in any part of the body not included in Stage I that is larger than 5 cm in diameter and/or has spread to regional lymph nodes.
   iv. Stage 4: Distant metastases are present at diagnosis. The primary tumor can be of any size or location.

7. Treatment: Multimodality approach

   a. Surgery

      i. Local control: Excision of primary tumor upfront whenever possible without causing major functional or cosmetic deficits. Wide resection of primary tumor with margin of normal tissue obviates the need for other local control measures in children with embryonal RMS.

      ii. Where feasible, pretreatment re-excision may be advisable to ensure complete tumor excision.

  - iii. However, when RMS occurs at sites where complete excision is not feasible or would result in an unacceptable loss of function, patients commence chemotherapy with incomplete excision or biopsy alone.
  - iv. Typically, complete excision is not considered for tumor arising in the orbit, most genitourinary sites, parameningeal sites, and some other sites.
  - v. Special anatomic sites requiring surgical assessment of retroperitoneal lymph nodes include paratesticular and extremity primary tumors larger than 5 cm (node sampling).
  - vi. In select cases, second-look surgery or delayed primary tumor excision may also be considered
  - vii. Patients that can have the tumor completely excised (Group1) generally have the most favorable outcome.
  - viii. For embryonal tumors, adequate surgical resection avoids the need for local irradiation.
  - ix. Tumors that are not completely excised (group II and III) typically receive local irradiation.

- b. Radiotherapy:
  - i. Local irradiation is indicated for most children and adolescents with RMS for patients with incompletely resected or gross residual (Group II and group III)
  - ii. Embryonal histology tumors irrespective of response to chemotherapy and for all patients with alveolar histology tumors
  - iii. Doses of 3,600–5,040 cGy generally used; dose depends on group (microscopic vs. gross disease), primary site, nodal involvement, histology, and whether second-look surgery performed.
  - iv. Timing of irradiation has varied from the fourth week of chemotherapy to the twelfth week of therapy, depending upon the tumor site.

- c. Chemotherapy:
  - i. Several chemotherapy agents have activity against RMS. Standard Chemotherapy regimen is vincristine, dactinomycin, and cyclophosphamide (VAC). Other active agents found to be useful in RMS include Temsirolimus, irinotecan, topotecan, doxorubicin, etoposide, and ifosfamide.
  - ii. Based upon the risk group chemotherapy regimens are modified
    1. Low-risk group represents about 30% of patient population and includes patients only with embryonal histology Group I or Group II or those with group III stage I. This group has a long-term survival of 93–94%. The approach to patients with low-risk RMS attempts to maintain excellent cure rates while decreasing the burden of care. Only vincristine and dactinomycin are used for the most favorable risk subset with cytoxan added for more extensive disease.

2. Intermediate risk Group: Represents 55% of patient's population and include patients with embryonal histology Group III, stage 2 and stage 3 as well as alveolar histology Group I-III and Group IV (under age 10 years)—This group has a long-term failure-free survival of 60–65%. VAC regimen remains the backbone for this group.
3. High-risk Group: Represents 15% of patient population and include Embryonal Group IV (>10 years) and Alveolar Group IV (all ages). This group has a long-term survival of 30%. Addition of different chemotherapy agents had little difference in overall outcome.

## Additional Notes

RMS has seen a gradual improvement to the modern overall survival rate of > 70%, even while there has been little change in the chemotherapy available [5]. Factors associated with worse survival are: age < 1-year-old, unfavorable site, bone/bone marrow involvement, and multiple metastatic sites [6]. Of the genetic markers identified, FOXO1 fusion status positive has been found to be the factor with greatest prognostic effect in predicting poor outcomes [1].

## Overall Long-term Outcome and Late Effects of Therapy

In view of the fact that the majority of patients with RMS are young and require for radiation therapy creates a concern. Orbital irradiation can cause cataracts, orbital hypoplasia, and pituitary insufficiency. Similarly, patients with bladder and prostate RMS have long-term issues with bladder dysfunction. Chemotherapy agents also increase the risk of gonadal toxicity, cardiotoxicity, and secondary malignancy. Further study of the genetic characteristics of RMS is leading to the development of potential targeted immunotherapy [7].

## Study Questions

Question 1: A 5-year-old child presents with symptoms of nasal obstruction and epistaxis, undergoes a biopsy, and is confirmed to have embryonal RMS. There are no distant metastatic sites involved. Based upon the IRS grouping system in which group will this child be designated:

a. Group I
b. Group II
c. Group III
d. Group IV

Answer to Question 1: (c.): Group III. A child who undergoes biopsy only as the initial surgical approach will be designated as Group III as per the IRS grouping system. Tumors located in the head and neck region are rarely amenable to complete surgical excision. Most of the time, only a biopsy is feasible, which can be done safely without functional or cosmetic deficits.

Question 2: A 3-year-old boy presents with progressive proptosis and esotropia. The tumor is biopsied, is confirmed to be embryonal RMS, and is confined to that site without regional node involvement or metastatic spread. Based upon the presentation, which risk group will this child be designated?

a. Low risk
b. Intermediate Risk
c. High risk
d. Very High risk

Answer to Question 2: (a.) Low risk. Per the IRS TNM staging, certain locations are considered favorable. These include the orbit, and non-parameningeal locations in the head and neck region. So, even though the child only had a biopsy, he will be considered to be group III and stage I. A child with group 3 and stage I is designated as low risk. Low-risk RMS carries a very good long-term survival.

Question 3: All the following are true for extremity RMS except:

a. Most extremity RMS are alveolar in histology.
b. Ipsilateral lymph node sampling is needed for accurate staging.
c. Chemotherapy is not indicated.
d. Radiation therapy is not indicated if RMS is embryonal histology and there is complete surgical excision with 2 cm margin of normal tissue.

Answer to Question 3: (c.) Chemotherapy is not indicated. RMS treatment usually involves multimodality-based treatment. However, if the tumor is able to be completely excised with adequate surrounding normal tissue, and it is embryonal histology without the involvement of the lymph nodes draining the region, local irradiation therapy can be avoided. Chemotherapy is indicated for all stages and groups of RMS. It effectively helps to control any micrometastasis which may occur and also prevents local recurrence. Proper ipsilateral lymph node sampling is critical for accurate staging purposes. Alveolar histology is seen in about 70% of extremity RMS and usually requires all 3 modalities of treatment: surgery, chemotherapy, and irradiation.

## References

1. Hibbits E, Chi YY, Hawkins DS, Barr FG, Bradley JA, Dasgupta R, Meyer WH, Rodeberg DA, Rugzinski ER, Spunt SL, Skapek SX, Wolden SL, Arndt CAS. Refinements of risk stratification for childhood rhabdomyosarcoma using FOX01 fusion status in addition to established clinical outcomes predictors: a report from the Children Oncology Group. Cancer Med. 2019;8(14):6437–48.

2. Dagher R, Helman L. Rhabdomyosarcoma: an overview. The Oncologist. 1999;4:34–44.
3. Pater LE, Breneman J, Wolden S, Donalds S. Rhabdomyosarcoma Review 2017: Children's Oncology Group. CA: Monrovia; 2017. https://www.qarc.org/COG/Rhabdomyosarcoma_pdf , Accessed 12 Nov 2021
4. Melempati S, Hawkins DS. Rhabdomyosarcoma: review of the Children's Oncology Group (COG) Soft-tissue sarcoma committee experience and rationale for current COG studies. Pediatr Blood Cancer. 2013;59(1):5–10.
5. Dasgupta R, Fuchs J, Rodeberg D. Rhabdomyosarcoma. Semin Pediatr Surg. 2016;25(5):276–83.
6. Oberlin O, Rey A, Lyden E, Bisogno G, Stevens MCG, Meyer WH, Carli M, Anderson JR. Prognostic factors in metastatic rhabdomyosarcomas: results of a pooled analysis from United States and European cooperative groups. J Clin Oncol. 2008;26(14):2384–9.
7. Chen C, Garcia HD, Scheer M, Henssen AG. Current and future treatment strategies for rhabdomyosarcoma. Front Oncol. 2019;9:1458. Frontiers Media, https://www.frontiersin.org/articles/10.3389/fonc.2019.01458/full. Accessed 11 Nov 2021

# Lymphomas

**Jagadeesh Ramdas and Marc S. Lessin**

**Key Points**

1) Lymphomas are the third most common malignancy of childhood and adolescence after leukemia and brain/central nervous system tumors.
2) Lymphomas are classified as Hodgkin and non-Hodgkin.
3) Lymphoma is a systemic disease. The treatment of lymphoma is medical. Chemotherapy is the mainstay of treatment with radiation therapy used sparingly.
4) The surgeon's role in the treatment of lymphoma is to obtain adequate tissue for diagnosis and immunohistochemical studies.
5) Ideally, a biopsy should be obtained under local anesthesia. Chest radiograph and CT should be obtained to determine if general anesthesia is contraindicated for mediastinal adenopathy.

Lymphomas are cancers that occur in children and adolescents as well as in adults. Lymphomas are classified as Hodgkin and non-Hodgkin lymphomas. Hodgkin lymphoma tends to occur more commonly in adolescence whereas non-Hodgkin lymphoma occurs in younger age groups. Lymphoma is considered a systemic disease much like leukemia. The primary treatment of lymphoma is medical with

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chemotherapy. Resections are generally not indicated. The surgeon's role is to provide tissue for diagnosis and intravenous access for chemotherapy. Special attention must be directed at the patient with large lymph nodes in the mediastinum which may preclude general anesthetics for biopsy procedures.

1. Epidemiology
    a. Incidence: Hodgkin lymphoma accounts for 6% of childhood cancers. Non-Hodgkin lymphoma accounts for 9% of all childhood cancers. Combined Hodgkin and Non-Hodgkin lymphoma (NHL) are the third most common malignancies in children and adolescents, with Hodgkin lymphoma being the most common cancer in people between the ages of 15 and 18 years.
    b. Etiology: Epstein–Barr virus (EBV) DNA-positive Hodgkin lymphoma is seen in younger patients less than 10 years of age. Patients with serologically confirmed infectious mononucleosis (either clinical or subclinical) have a 3–5-fold increased risk for developing EBV-positive Hodgkin lymphoma.
    c. Inherited and acquired immunodeficiency is a strong risk factor for developing Non-Hodgkin lymphoma during childhood. Most common Inherited immunodeficiency at risk include Wescott–Aldrich syndrome (WAS), severe combined immunodeficiency (SCID), X-line lympho-proliferative disease (XLP), autoimmune lymphoproliferative syndrome (ALPS), common variable immunodeficiency (CVID), X-linked hyperimmunoglobulin M syndrome, ataxia telangiectasia.
2. Pathophysiology:
    a. Classification Schemes:

        Hodgkin Lymphoma

        i. Lymphocyte predominant which is the least severe subtype.
        ii. Mixed cellularity which is of intermediate severity.
        iii. Nodular sclerosing which is of intermediate severity and is the most common variation in the adolescent population.
        iv. Lymphocyte depletion is the most severe subtype.

        Non-Hodgkin lymphoma classification is based on cell of origin, primarily whether B-cell or T-cell based:

        i. B-cell lymphoma often associated with Burkitt lymphoma & Burkitt like lymphoma –usually involves abdomen, head, neck, bone marrow, and CNS
        ii. Lymphoblastic lymphoma of B & T-cell origin—usually involves mediastinum, lymph nodes, CNS, bone marrow, and abdomen
        iii. Diffuse large B-cell lymphoma—involves mediastinum and abdomen
        iv. Anaplastic large cell lymphoma of B & T-cell origin—involves skin, mediastinum, liver, spleen, and abdomen

## Hodgkin Lymphoma Staging

I. Single lymphatic site
II. Two or more lymphatic sites on the same side of the diaphragm
III. Disease involving above and below the diaphragm. Involvement of the spleen if contiguous to an involved lymph node region
IV. Multifocal involvement of one or more extralymphatic organs or tissues with or without lymph node involvement, or isolated extralymphatic organ involvement with distant nodal involvement

B Symptoms: Unintentional weight loss of > 10% in 6 months, unexplained fevers > 38°C for more than 3 days, or drenching night sweats.

B symptoms are associated with a worse prognosis.

Bulky disease—Widening of the mediastinum by more than one third or the presence of a nodal mass with a maximal diameter >10 cm

## Non-Hodgkin Lymphoma Staging

Stage 1—Single tumor (extra nodal) or single anatomic area (nodal).
Stage II—Single tumor with regional nodal involvement
Two or more nodal areas on the same side of the diaphragm
Stage III—Two single tumors on opposed sides of the diaphragm

- Two or more nodal areas above and below the diaphragm
- All of the primary intrathoracic tumors (mediastinal, pleural, thymic)
- All extensive primary intrabdominal disease
- All paraspinal or epidural tumors

Stage IV—Any of the above with initial CNS and/or bone marrow involvement (<25% malignant cells)

## Pathology: Non-Hodgkin Lymphoma

NHL is a diverse collection of malignant neoplasms of lymphoid cell origin. They are heterogeneous in their pathology, clinical features, and responsiveness to therapy. Most common ones seen in children and young adults are:

1. Burkitt and Burkitt like lymphoma- Small non-cleaved cell lymphomas. Diffuse homogenous proliferation of intermediate-sized cells larger than lymphocytes. Burkitt lymphoma has an extremely high proliferative rate (doubling time of 24 h) and a *starry sky* pattern of reactive macrophages—*Immunophenotype*- positive for CD19, CD20, CD22, CD10 and cell surface immunoglobulin.

Cytogenetics- translocations involving C-MYC oncogene locus on choromosome8, (8:14) q24; q32) translocations.

2. Lymphoblastic Lymphoma—Immature, blast-like appearance with fine chromatin, inconspicuous or absent nucleoli and scanty cytoplasm. Cytologically the neoplastic cells are indistinguishable from those seen on precursor B or T cell acute lymphoblastic leukemia. Immunophenotype positive for CD19, TdT, CD2
3. Diffuse large B cell lymphoma—Large noncleaved cells, large cleaved cells, polylobated cells, and immunoblastic cells. Mature B cell immunophenotype expression of CD20, CD22, and CD79a.
4. Anaplastic large cell lymphoma—large pleomorphic cells, multinucleated cells or cells with eccentric horse-shaped nuclei and abundant basophilic cytoplasm. Strong CD30 expression. Characteristic translocation t (2:5) (p23.q35)

## Pathology: Hodgkin Lymphoma

The hallmark of Hodgkin lymphoma is the Reed Sternberg cell. These are multinucleated cells with two prominent nucleoli which have an appearance described as "owl's eyes." They usually express cluster of differentiation (CD) 15 and CD30 antigens.

5. Diagnosis

   a. Symptoms: The most common presentation in Hodgkin lymphoma is persistent and painless enlarged lymph nodes typically in the neck and supraclavicular region. The nodes are described as "rubbery" and often multiple. Presentation is usually indolent and slowly progressing. History of "B" symptoms should be obtained including fevers, night sweats, or any changes in weight. B symptoms are seen in about 20–25% of patients.

      Some patients may have significant pruritus at the time of presentation. Patients with large anterior mediastinal mass may present with cough or shortness of breath and may show superior vena cava compression syndrome.

      The vast majority of NHL (Non-Hodgkin lymphoma) usually present with advanced disease. Endemic Burkitt lymphoma seen more in equatorial Africa will commonly present with head and neck involvement, especially jaw involvement. Sporadic form for Burkitt lymphoma seen more commonly in North America and Europe presents with abdominal involvement. Some patients occasionally present with small bowel pathology. Lymphomas have been associated with lead point intussusceptions, bowel perforation, and bowel obstruction.

      Childhood lymphoblastic lymphoma commonly presents with a supradiaphragmatic mass usually either intrathoracic and/or mediastinal. Presentations include various forms of respiratory distress and signs and symptoms associated with superior vena caval syndrome. Often presents with pleural effusion and pericardial effusion

Diffuse Large B cell lymphoma also presents similar to other forms of small non-cleaved cells lymphoma. Advanced DLBCL presents with an abdominal primary lesion and to a lesser extent, a primary lesion in the mediastinum.

Anaplastic large cell lymphoma (ALCL) has a different clinical presentation resulting in two categories. Systemic and primary cutaneous ALCL. Systemic ALCL has more extranodal disease involving skin, soft tissue, and bone. Primary ALCL is more confined to the skin.

b. Examination: A complete physical examination with special attention to the superficial lymph node areas including the neck, supraclavicular region axilla, epitrochlear nodes, and groins. Abdominal examination should be performed to determine if abdominal mass or organomegaly is present.
c. Laboratory Tests: Blood work should be obtained including complete blood count, C-reactive protein, erythrocyte sedimentation rate, hepatic function testing, electrolytes including blood urea nitrogen and creatinine, lactate dehydrogenase, uric acid, and alkaline phosphatase. Bone marrow biopsy and aspirate should be obtained. Lumbar puncture and checking cerebrospinal fluid for involvement is also needed especially for Non-Hodgkin lymphoma for proper staging.
d. Radiographic Studies: Chest X-ray should be obtained to ascertain if mediastinal adenopathy is present. CT scans of the neck, chest, abdomen, and pelvis should be obtained. CT PET scan is also commonly used with fluorodeoxyglucose (FDG) to determine the extent of disease. FDG is taken up by hypermetabolic areas. Functional imaging is particularly important in Hodgkin lymphoma as residual masses are seen at the end of the treatment and the anatomical distinction of scar from active tumor is not possible. Radiological documentation, especially a PET negative scan documents a rapid early anatomical response to chemotherapy and has been identified as a very important prognostic factor to decide about subsequent chemo regimen as well as the need for irradiation therapy in Hodgkin lymphoma.

6. Treatment

a. Initial Measures: The surgeon's role is to obtain adequate tissue for diagnostic purposes and to perform immunohistochemistry studies.
b. Preoperative Preparation: Mediastinal involvement which is present in three-fourths needs to be considered in all suspected lymphomas. In patients that present only with a mediastinal mass, a biopsy target that can be accessed under local anesthesia should be sought. Pleural fluid may be aspirated by thoracentesis if pleural effusion is present. Bone marrow aspirate and biopsy may be diagnostic. In patient with large mediastinal involvement, general anesthetic is contraindicated. There are multiple reports of airway and/or cardiovascular collapse after undergoing induction of general anesthesia with a paralytic agent. Patients at risk may present with respiratory distress, orthopnea, or inability to lie in the supine position. In these patients, the diameter of airway can be assessed on CT scan. Perger reported that patients with more

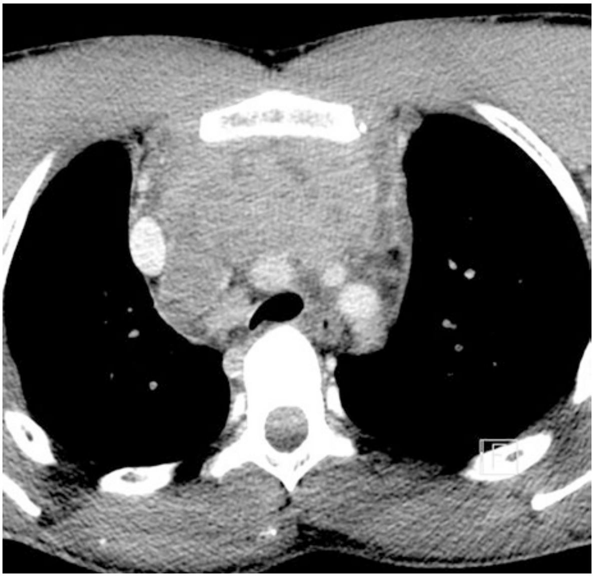

**Fig. 1** CT chest showing mediastinal disease which would be concerning for a general anesthetic with a paralyzing agent (Source: Marc Lessin, M.D.)

than a 50% reduction in tracheal area or peak expiratory flow rate are at risk for cardiovascular collapse during induction of anesthesia requiring paralytic agents. In cases where no tissue is accessible under local anesthetic, it may be necessary to perform mantle radiation of the mediastinum before biopsy which makes subsequent diagnostic studies difficult (Fig. 1).

c. Operative Treatment: Most commonly, the surgeon is called upon to provide tissue for diagnosis. Adequate tissue is required for accurate diagnosis, staging, and immunohistochemical studies. Needle aspiration or core needle biopsy is often inadequate for tissue diagnosis. Staging biopsies including splenectomy is no longer considered necessary or appropriate. Bone marrow biopsy and lumbar puncture are often performed in conjunction with biopsy for staging purposes. Intrathecal chemotherapy may be combined with lumbar puncture if indicated.

   Occasionally, the presentation is small bowel obstruction or intussusception. In these cases, segmental small bowel obstruction with anastomosis may be necessary. This will satisfy the dual purpose of relieving the bowel obstruction and providing tissue for diagnosis.

   Mucosa-associated lymphoid tumor is a rare entity that is diagnosed on esophagogastroduodenoscopy (EGD). The tumor is associated with Helicobacter pylori infection. Treatment is directed at the treatment for the Helicobacter.

d. Treatment of pediatric Hodgkin lymphoma—Primary treatment of Hodgkin lymphoma is chemotherapy and all pediatric patients receive chemotherapy. Various chemotherapy regimens have been studied by different cooperative groups and various components of chemotherapy regimen, individual dosage, frequency, and intensity, and the total number of cycles are modified based upon the initial stage of the lymphoma as well as based upon the radiological early response seen after initial rounds of chemotherapy. ABVD, ABVE -PC, BEACOPP, OEPA, COPDAC, COPP-ABV, VAMP, DBVE, Stanford V, OPPA are some of the regimens which have been used in the management of pediatric Hodgkin lymphoma across various cooperative groups. Radiation therapy is also used in certain situations and remains a standard of care. However, in children, the goal of eliminating radiation therapy for patients achieving a complete response to chemotherapy stems from the premise that eliminating radiation will lead to a decrease in radiation-related late effects. Radiation fields used in treating Hodgkin lymphoma have also evolved from using total nodal irradiation to mantle field and then to involved field and now to involved site irradiation. Using irradiation therapy only for large bulky disease as well as those with initial inadequate response to chemotherapy has also been used.

Since Hodgkin lymphoma has a high overall cure rate, risk stratification strategy to maximize treatment efficacy, and at the same time minimize immediate as well as late toxicities are being adopted.

Treatment strategies for Non-Hodgkin lymphoma—Since NHL are heterogeneous, chemotherapy treatments and regimens used in different types of NHL are also variable. Lymphoblastic lymphomas are primarily treated based upon protocol adopted for acute lymphoblastic leukemia, whereas mature B cell lymphoma like large B cell and Burkitt lymphoma are treated with a shorter duration but more intense chemotherapy regimens which uses multiple drugs over a shorter period of time. Treatment risk stratification is based upon stage, initial LDH values, CNS and bone marrow involvement.

## Additional Notes

Emergencies in patients with newly diagnosed lymphoma—Children with lymphoma can have life-threatening findings such as respiratory distress and superior vena cava syndrome (SVC syndrome) caused by a mediastinal tumor, bowel obstruction from large abdominal masses, cranial nerve palsies, and paraplegia in cases of spinal cord involvement. Invasive diagnostic measures, especially those requiring anesthesia can create emergencies for children with lymphoma.

Diagnostic procedures may need to be done with local anesthesia alone or with least invasive procedure for getting diagnostic tissue. Patients with impending cord compression may need surgical decompression with laminectomy, steroids, and irradiation therapy.

Metabolic derangements can occur with rapid tumor cell breakdown leading to the development of acute tumor lysis syndrome (ATLS). ATLS is characterized by various combinations of hyperuricemia, hyperkalemia, hyperphosphatemia with or without hypocalcemia. If untreated this can rapidly lead to seizures, cardiac arrhythmias, renal failure, and death. Patients with Non-Hodgkin lymphoma (NHL) are at higher risk, particularly those with advanced stage, Burkitt lymphoma, and lymphoblastic lymphoma with high LDH values. ATLS needs to be managed with vigorous hydration, alkalinization, and administration of allopurinol as well as in some cases Recombinant urate oxidase agents like Rasburicase.

## Study Questions

1) Which is an accepted concern for Hodgkin lymphoma survivors?

   a. Learning disability and neurocognitive dysfunction
   b. Avascular necrosis of the bone
   c. Second malignant neoplasm
   d. Increased risk of Infections

   Answer to Question 1: c. Hodgkin Lymphoma typically does not affect the central nervous system and they do not get any CNS-directed therapy. Risk of avascular necrosis of the bone is also limited due to limited long-term steroid exposure. Splenectomy is not done anymore as part of staging, so that patients are not permanently immunocompromised. The development of second malignant neoplasm (SMN) in Hodgkin lymphoma survivors is a major area of concern. The overall risk of developing an SMN is approximately 2% at 5 years from completion of treatment, 5% at 10 years and 10% at 15 years of treatment. There appears to be a singularly heightened incidence of breast cancer in young women who have received standard dose radiotherapy (3500–4400 cGY) to their mediastinum/chest; the incidence increases with time and may approach 20% at 20–25 years from the time of treatment. Early initiation of surveillance screening with mammogram and MRI by 25 years are recommended for Hodgkin lymphoma survivors

2) 8-year-old boy presents to your office with abdominal pain, constipation, with a history of anuria for the last 24 h. On examination, you notice a mass across the lower abdomen with few scattered enlarged lymph nodes. You get an abdominal ultrasound which shows a diffuse retroperitoneal mass along with dilated ureters and evidence of bilateral hydronephrosis. Which is the best approach

   a. Arrange for a biopsy in the OR next day
   b. Start hyperhydration immediately
   c. Placement of external nephrostomy stent as soon as possible
   d. Give steroids

Answer to Question 2: (c); Childhood non-Hodgkin lymphomas are one of the extremely fast-growing neoplasms; The doubling time of Burkitt lymphoma can range from 12 h to a few days. Depending upon the anatomic location of the tumors, this rapid growing time can result in severe symptoms due to the sheer bulk of the disease. In view of the obstructive uropathy before initiating any hyperhydration, it is important to establish a urinary outlet to preserve renal function. Similarly, small bowel tumors may precipitate an acute intestinal obstruction or perforation, epidural masses can cause spinal cord compression and paraplegia; mediastinal tumors can cause airway compromise due to tracheal compression.

3) Fifteen-year-old male with history of fever, night sweats, and weight loss. Complains of shortness of breath and difficulty lying flat. Chest x-rays show massive mediastinal lymphadenopathy. A thorough physical examination shows no palpable lymphadenopathy. CT-PET scan shows uptake in the chest and abdomen. Bone marrow biopsy is negative for tumor. Biopsy of the mediastinal disease is requested. The best approach is:

   a. Thoracoscopic biopsy of mediastinal lymph nodes.
   b. Core needle biopsy of the mediastinal nodes.
   c. Neck exploration under local anesthesia for node biopsy.
   d. Preoperative radiation therapy with shielding to avoid exposure of selected nodes.

Answer to Question 3: (d). The large mediastinal mass presents as a significant problem in the diagnosis of lymphoma. There are numerous case reports of sudden death in patients with large mediastinal disease. Patients undergoing general anesthetic are at risk for cardiovascular collapse after receiving paralytic agents for general anesthetic. Intubation will not rectify the problem. The etiology is believed to be combination of airway collapse due to medical paralysis of the voluntary muscles of respiration and/or collapse of the vena cava preventing blood return to the heart. In these situations, general anesthesia must be avoided. Every attempt should be made to determine if biopsy can be obtained under local anesthesia from a superficial lymph node, pleural effusion, or bone marrow. Core needle biopsies are considered inadequate for tissue typing although a recent paper out of England cites success with this approach. In the case when no other option for tissue diagnosis other than the mediastinum, preoperative radiation of chemotherapy should be started to shrink the mediastinal involvement. Radiation provides the option of shielding target nodes whereas systemic chemotherapy will change the histologic appearance of all involved tissue.

## Further Reading

Acker SN, Linton J, Tan GM, Garrington TP, et al. A multidisciplinary approach to the management of anterior mediastinal masses in children. J Pediatr Surg. 2015;50(5):875–8.

Alexander S, Fernando A. (2015) Pediatric lymphoma In David G Nathan; Stuart H Orkin; Frank A Oski Nathan and Oskis: hematology and oncology of infancy and childhood, 1626-1671, Philadelphia, PA: Elsevier

Malik R, Mullassery D, Kleine-Brueggeny M, Atra A, et al. Anterior mediastinal masses – a multidisciplinary pathway for safe diagnostic procedures. J Pediatr Surg. 2019;54(2):251–4.
Nachman J. Hodgkin lymphoma. In: Carroll WL, Finlay JL, editors. Cancer in children and adolescents. Sudbury, MA: Jones and Bartlett; 2010. p. 217–41.
Perger L, Lee EY, Shamberger RC. Management of children and adolescents with a critical airway due to compression by anterior mediastinal mass. J Pediatr Surg. 2008;43(11):1990–7.

# Part IX
# Surgical Procedures

# Parenteral Access

**Kody L. Moore and Christopher P. Coppola**

**Key Points**

1. Intravenous access in a child can be one of the most difficult procedures, particularly in small children with small veins hidden by subcutaneous fat.
2. In an emergency, if no access is obtained within three attempts or 90 seconds, an intraosseous line provides rapid access through which a child can receive fluids, medications, and transfusions.
3. Whenever possible, existing intravenous lines should be preserved with antibiotics, thrombolytic therapy, or antiseptic locks as there are a limited number of veins available in a child.

1. Percutaneous approach can be used for peripheral and central veins. For central veins, sonography before and during line placement reduces complications and increases the rate of success. Check coagulation function before line attempt. Most children require anesthesia for safe line placement, and success rate will be higher in a patient who is still.
    a. Peripheral: for most therapy, peripheral access will suffice. Use of peripheral IV catheters reduces the risk of line sepsis and thrombosis. Vein is located, tourniquet is applied, skin is prepped, and catheter is inserted at a low angle with skin, parallel with vein, attempting to thread the needle into the vein, then slide catheter off needle. Catheter is flushed, secured with an occlusive dressing, and for young patients, the limb is secured with a padded immobilizer. In small children, the scalp veins can be used.

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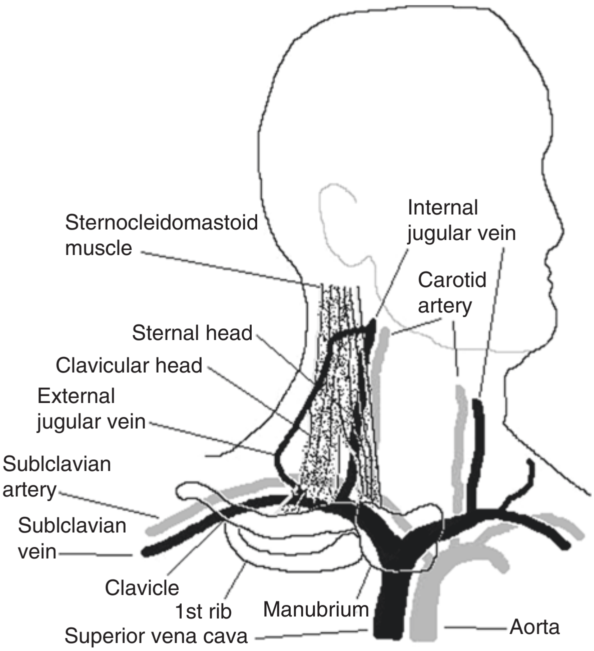
**Fig. 1** The jugular vein, subclavian vein, and associated anatomic landmarks (Source: Christopher Coppola)


Sternocleidomastoid muscle
Internal jugular vein
Carotid artery
Sternal head
Clavicular head
External jugular vein
Sublclavian artery
Sublclavian vein
Clavicle
1st rib
Manubrium
Superior vena cava
Aorta

b. Jugular (Fig. 1): Patient is positioned supine with 15–30 degrees Trendelenburg to distend veins and avoid air embolus. Head is turned away from the side being attempted and a rolled towel under the shoulders helps extend the neck. Sonography is used to locate the jugular vein and carotid artery. Vein can be approached from a vector that starts posterior to the sternocleidomastoid muscle, between the clavicular and sternal heads of the muscle, or anteriorly, through the belly of the muscle itself. Skin is prepped and sterile technique is used. Vein is accessed with a needle and syringe using sonography and palpation to avoid the carotid artery. Seldinger technique is used to place a wire in the vein, advance a catheter over the wire, and then the wire is removed. It is verified that the catheter withdraws blood and flushes saline easily. Catheter is secured to skin with sutures and an occlusive dressing. Chest x-ray is used to verify that the tip of the catheter is at the cavoatrial junction and there is no pneumothorax.

c. Subclavian: Patient is positioned supine, with Trendelenburg, and a roll between the shoulder blades. Sonography is used to identify the subclavian vein. Sonography of the subclavian vein is more difficult than the jugular vein due to the clavicle. There are supra-clavicular and infra-clavicular windows. Shoulder and neck are prepped and alcohol and sterile technique used. Vein is accessed with a needle and syringe by entering skin lateral to the site where the clavicular head of the pectoralis muscle attaches to the curve in the clavicle. Needle is kept parallel to the floor, the tip is allowed to contact the clavicle, and then is redirected between the clavicle and the first rib until the subclavian vein is entered and venous blood can be withdrawn. Aim the tip of

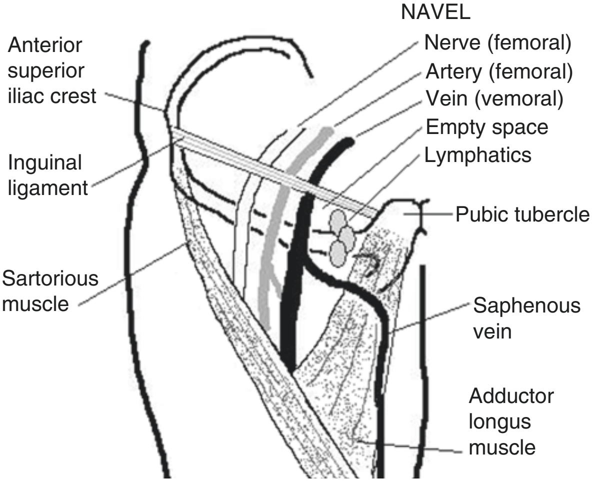


**Fig. 2** Femoral artery, vein, and associated landmarks (Source: Christopher Coppola)

the needle at the manubrium or larynx. Avoid directing the needle toward the floor as this will increase the chance of pneumothorax. Using the Seldinger technique, the catheter is placed, ensuring that the wire is completely removed. Fluoroscopy during procedure verifies the wire is directed to the superior vena cava and not to the opposite arm or up into the neck. An 11-blade scalpel and a dilator are used to widen the entry so the catheter can enter. Catheter is secured and dressed, then chest X-ray is performed to verify placement in the superior vena cava and ensure there is no pneumothorax.

d. Femoral (Fig. 2): Patient is placed in supine position. Femoral vein is located with sonography and note is taken of the femoral artery, lateral to the vein. After sterile prep and drape, vein is accessed with a needle and syringe, at a downward angle with needle parallel to the vein. When vein is entered, using Seldinger technique a wire is inserted into vein and is used to advance a catheter into the femoral vein. The wire is removed. It is verified that the catheter can withdraw and flush, and it is secured with a suture and occlusive dressing.

2. Cut down

   a. Jugular:

      i. Technique: Patient is positioned for percutaneous access. The space between the sternal and clavicular heads of the sternocleidomastoid muscle is palpated. A transverse incision is made a finger breadth above the clavicle. The subcutaneous tissue and platysma are separated, and wound is deepened until carotid sheath is encountered. The internal jugular vein is identified. It is carefully separated from the carotid artery and the vagus nerve. Proximal and distal control of the vein is obtained by circumferential dissection and encircling vein with 4-0 absorbable ties above and below the planned point of vein entry. The distal vein is ligated. Venotomy is created. Catheter is inserted and advanced until the tip lies within the superior vena cava. The proximal ligature is tied down ensuring that it does not crush the catheter. After verifying that the catheter withdraws blood and flushes saline, the wound is closed in layers of absorbable suture. The catheter is secured to skin and covered with an occlusive dressing.

    ii. Alternatives:

        1. The internal jugular does not necessarily need to be ligated during catheter placement. A purse string of 4-0 polypropylene suture can be used to secure the edge of the venotomy, which is then tied down around the catheter, preserving jugular flow from the head.
        2. A branch of the internal jugular, the facial vein, can be used as the entry point for the catheter, and the internal jugular will not need to be ligated.
        3. In larger children, the external jugular can be used for placement. After proximal and distal control is obtained, catheter is inserted via venotomy and must be guided into external jugular, which leads to the subclavian, and then enters the superior vena cava, via the innominate vein if on the left side.

  b. Femoral: Cutdown is usually performed parallel and just inferior to the inguinal ligament, but a vertical incision is an alternative. From lateral to medial, the orientation of structures in the femoral triangle are Nerve, Artery, Vein, Empty space (occupied by fat), and Lymphatics (NAVEL). The femoral vein is identified, dissected out circumferentially, and secured with ties proximally and distally. Catheter is inserted via venotomy. Flow in the femoral vein is preserved.
  c. Saphenous vein runs from ankle to groin, on the medial surface of both legs. It is found easiest at the ankle, where a transverse incision is made a finger breadth superior and anterior to the medial malleolus. The vein is identified, isolated with proximal and distal control, distal vein is ligated, and catheter is inserted through venotomy, then proximal ligature is tied down to secure vein around catheter. Saphenous vein can also be found two finger breadths inferior and posterior to the femoral vein at the inguinal ligament.

3. Peripherally inserted central catheter (PICC): A PICC line offers the advantages of a central line, since the tip lies in the superior vena cava, but with fewer mechanical and infectious complications. The best vein for PICC insertion is the basilic vein above the antecubital fossa. The vein is identified with the help of sonography. Distance from entry site to the upper chest along path of venous return is measured to estimate length of catheter that will be required. Tourniquet is applied above entry site. Vein can be accessed by percutaneous technique or cutdown. Vein is accessed by a breakaway needle that splits in half, or a short catheter, large enough to pass the PICC line, depending on which type of kit is available. PICC is inserted through this device into vein. Tourniquet is removed. PICC line is advanced until the tip reaches the superior vena cava

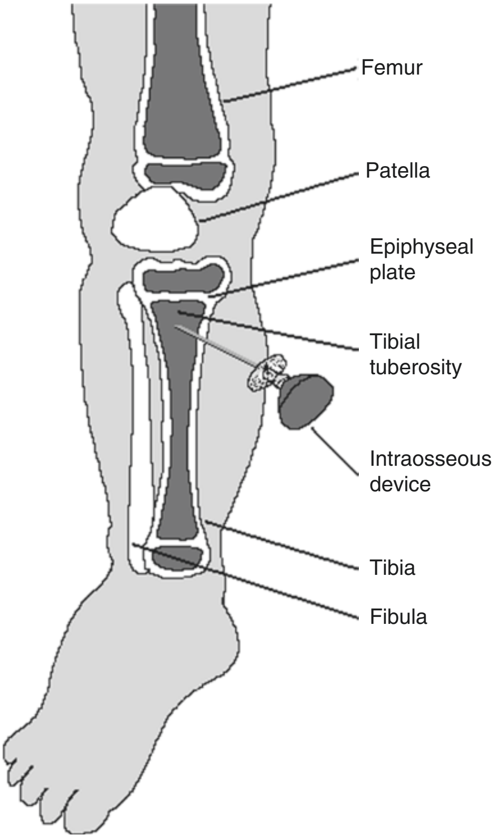


**Fig. 3** Intraosseous line insertion into the proximal tibia (Source: Christopher Coppola)

4. Intraosseous (IO) (Fig. 3)
   a. Indications and use: An intraosseous line is a useful rescue when an intravenous line cannot be obtained. In an emergency, an intraosseous line should be placed when three attempts have been made or 90 seconds have passed without progress. Line can be used for IV fluid, medications, transfusion, and phlebotomy. It can be started easily on children up to age 6 years old, above that age, entry into bone can be difficult without specialized devices.
   b. Devices:
      i. Hand-driven device with detachable handle: Cook Intraosseous, either threaded or smooth needles, (Cook Medical, Bloomington, IN); Jamshedi 15g, (CareFusion, San Diego, CA)
      ii. Battery-powered drill: EZ-IO, (Vidacare, San Antonio, TX)
      iii. Spring-loaded needle: "Bone Injection Gun" (BIG), (Waismed Ltd., New York, NY)
      iv. Sternal intraosseous insertion device (age 12 years and above): "First Access for Shock and Trauma 1" (Fast 1), (Pyng Medical Corp., Richmond, British Columbia, Canada)

c. Entry site

    i. Proximal tibia is the preferred insertion site and works in children and adults. Medial surface of the tibia is flat, and overlying tissue is thin. This allows for palpation of landmarks and minimum tissue to traverse before bone. From the tibial tuberosity, measure 1 cm distal and 1 cm medial (2 cm for children > 6 years), finding a broad flat area where skin is thin over the tibia.
    ii. Sternal placement of an intraosseous needle requires a commercially available device which can be used in patients 12 years old and older. The device comes with a positioning patch which guides needle entry approximately 2 cm below the sternal notch. The catheter comes with a small extension set, protective dressing, and specialized removal tool which is kept with the patient.
    iii. Alternate placement sites used when primary sites are unavailable due to trauma or previous attempt include the distal tibia on medial aspect, distal femur on anterior aspect, proximal humerus on antero-medial aspect, iliac crest, and others. In general, choose a site without overlying neurovascular structures to be injured, in the portion of bone with marrow cavity, and with minimum thickness of subcutaneous tissue.
    iv. Contraindications (most are site-specific, and an alternate site can be chosen):

        1. Fracture of bone
        2. Previous attempt in same bone
        3. Vascular injury to limb
        4. Disease of target bone (tumor, osteoporosis, osteogenesis imperfecta)
        5. Infection or tissue deficit of soft tissue at target site

    v. Complications: Intraosseous lines are used for expedient access when prompt intravenous access is unavailable. Although they may endure for up to 96 hr, they should be replaced with intravenous access as soon as it is safely and accurately available.

        1. Erroneous placement outside bone or completely through bone: this will cause extravasation
        2. Extravasation around needle: this increases the longer the needle is used
        3. Compartment syndrome from extravasation
        4. Infection
        5. Dislodgement of needle
        6. Occlusion of needle by bending, bone, or clot
        7. Hematoma
        8. Air or fat embolus
        9. Pneumothorax, hemothorax, or mediastinitis from through and through penetration of sternum

5. Creating a tunnel under the skin for passage of the catheter before it enters the vein provides protection against infection. Also, the catheter can be secured to the subcutaneous tissue by cuffs of fibrous material into which the body grows and attaches. Tunneled catheters are safer for outpatients as there is less chance of catheter dislodgement and hemorrhage. Tunneled catheters are placed in the operating room or interventional radiology suite with anesthetic. Catheter tip is positioned at the junction of right atrium and superior vena cava, confirmed by chest X-ray. Catheter is accessed with aseptic technique to avoid contamination. Heparin 100 U/mL is left indwelling in the catheter to avoid thrombosis, unless contraindicated. In some scenarios, use of an indwelling ethanol lock can reduce infection.

   a. Externalized catheters enter the skin at one location, traverse the subcutaneous space, then enter a vein at a different location. Examples are Broviac and Hickman catheters. (C.R. Bard Inc., Murray Hill, NJ). Cuff is positioned under the skin for catheter fixation.
   b. Completely implanted catheters have a reservoir under the skin, and a catheter that traverses the subcutaneous space before entering the vein. For infusion or phlebotomy, the reservoir is accessed through the skin using a Huber needle, which is a non-coring needle that will not create leaking voids in the silicone surface of the port. Examples are Mediport and Port-a-cath (Smiths Medical, St. Paul, MN). The best location for the reservoir is on the surface of the pectoralis muscle fascia. The skin in the midline chest can be too thin and will erode with repeated use. The breast should be avoided because incision creates future scarring in breast, breast tissue thickness makes access of the port difficult, and when the patient stands, the weight of the breast can pull the port inferiorly and retract the catheter tip out of proper position. Care is taken to ensure that the catheter is securely fastened to the spout exiting the port to avoid leakage. The port should be secured to the pectoralis fascia in at least two locations to avoid port migration and flipping. Port placement will leave a larger scar than an externalized catheter and will require a second operation to remove. Ports give the advantage of being more difficult to break or dislodge. Patients will be able to swim and shower when the port is not accessed.

6. Dialysis

   a. Hemodialysis: Various catheter options exist included tunneled vs. percutaneous, and a range in sizes of catheter diameter and length to cover pediatric patients across changes in age and size. The catheters have two lumens, corresponding to the need to simultaneously withdraw blood from the patient and return blood after it has passed through the dialyzer. The tips of the two lumens are staggered to reduce the recirculation of dialyzed blood directly back into the catheter. Catheter is positioned so that the tips of both lumens are clear with good withdrawal of blood and easy flushing of saline. Heparinized saline is left indwelling in both lumens of the catheter. The concentration of heparin used can vary between institutions; one example is 1000 U/mL. Before flushing the catheter, the indwelling heparin must be withdrawn and discarded to avoid infusing the heparin and causing bleeding.

  i. Percutaneous hemodialysis catheters are used when the need is urgent and the catheter is intended to be used temporarily, while the child is an inpatient, and removed when the child recovers renal function, or a more durable access for dialysis is provided. The best access for percutaneous hemodialysis is the right internal jugular vein because it provides a straight, short path to the right atrium without flexing of the catheter which can reduce flow rate.
  ii. A tunneled hemodialysis catheter is placed when a patient is intended to have the catheter for a prolonged period and have the catheter while they are an outpatient. The routes most used are the right internal jugular vein and the right subclavian vein because the catheter tip will descend the superior vena cava in a straight superior to inferior orientation, avoiding occlusion caused when the catheter is pressed up against the inside surface of the superior vena cava.

b. Peritoneal dialysis is preferred by most families because therapy can be provided in the home. It also avoids the fluctuations in blood pressure that occur during hemodialysis

  i. Specialized peritoneal dialysis catheters called Tenckhoff catheters are used. There are different sizes available for different ages. Catheters have multiple holes to allow for the efficient flow of dialysate and peritoneal fluid, and to reduce the incidence of catheter occlusion from adhesions. There is a portion of the catheter intended to lie in a subcutaneous tract, and one or multiple fibrous cuffs to encourage tissue ingrowth for fixation. The catheters are pre-curved to assist with the creation of a subcutaneous tract and creation of a region of the peritoneum that serves as a reservoir for dialysate. Examples are the Quinton Curl Cath and Argyle Swan Neck Cath (Covidien, Mansfield, MA).
  ii. Catheter is inserted sterilely in the operating room. A supraumbilical incision is made to gain entry to the peritoneal cavity. The catheter tip is directed downward, to lie in the anterior pelvis. The peritoneum and midline fascia are closed with a purse-string suture, attempting to create a water-tight seal. The external portion of the catheter is tunneled out to an incision in the right abdomen, where it is secured with an occlusive dressing. The fibrous cuff or cuffs of the catheter must be under the skin.
  iii. Omentectomy can be performed at the time of peritoneal dialysis catheter placement to prevent future occlusion of the catheter. If omentectomy was not performed at the time of initial placement, it can be performed to salvage an occluded catheter.
  iv. When the need is urgent, the peritoneal dialysis catheter can be used immediately, but there will often be leakage of dialysis from the two inci-

sions. When possible, peritoneal dialysis should be delayed at least 2 weeks to allow the body to create a tight seal around the catheter.

7. Arterial access is often needed in critically ill patients and during operations with anticipated fluctuations in blood pressure. They can be used to continuously monitor blood pressure and to withdraw blood specimens.

    a. Placement: Arterial site is selected. Skin is prepped. An arterial catheter is selected: size should be 18–24 gauge depending on the age of patient. Either intravenous catheters can be used, or specialized kits which include a wire to guide advancement of catheter into artery. While palpating artery, catheter is inserted into skin at a 45-degree angle until a flash of blood is seen. Needle angle is lowered, and needle is slightly advanced to thread into the lumen of artery. If a wire is present, it can be advanced into artery at this time. Catheter is advanced off needle into the artery and the needle and wire are removed. There should be a pulsatile flow of blood from the catheter and an arterial wave form when connected to a monitor. Catheter is secured with a suture and an occlusive dressing is applied. When percutaneous placement attempt is unsuccessful, open cutdown to visualize artery during catheter placement can be performed.

    b. Upper extremity sites

        i. Radial artery: Before placing a radial arterial line, Allen's test is performed. Radial and ulnar arteries are compressed, hand is squeezed to blanch skin, ulnar artery pressure is released, and it is verified that color returns to the hand within 5 s. If test is abnormal, consider another site. Hand and wrist are stabilized on an arm board with dorsiflexion of the wrist for placement. During radial artery line use, the hand and fingers are observed for signs of thrombosis.
        ii. Axillary artery: This site is a secondary site when radial arteries are not available or palpable. It can also be used for catheter-based interventions. When axillary hair is present, it should be trimmed to allow for more effective skin prep and application of an occlusive dressing. There is more motion possible in the axilla than the wrist so patients should be stabilized and sedated when necessary while an axillary artery catheter is in place.

    c. Lower extremity

        i. Femoral artery: Artery crosses the inguinal ligament halfway between the anterior superior iliac spine and the pubic tubercle. A long enough catheter should be selected to traverse the subcutaneous fat and so that tip does not become dislodged from the artery with patient movement. Length varies with age; five and eight cm lengths are available. After femoral artery catheter placement, the leg needs to be monitored for signs of thrombosis and ischemia.

ii. Dorsalis pedis artery: This artery runs over the dorsum of the foot, toward the medial aspect. It can usually be located at the crease on the anterior ankle. Foot is dorsiflexed and secured. Catheter should enter artery below ankle crease, so it is not occluded with foot flexion.
iii. Posterior tibial artery: This artery runs posterior to the medial malleolus of the ankle. It is usually encountered deeper than expected by palpation of the pulse.

## Additional Notes

Vascular access is a crucial necessity in the treatment of children (Fig. 4). When vascular access is needed, one of the more commonly performed procedures by pediatric surgeons is the placement of the above-described access devices. Despite the high prevalence of these line placements, they are not without risk. There is a need for careful consideration in preserving them and limiting the need for multiple replacements. Every child is born with a set number of vessels suitable for an access point. The vessels that are present at birth will be the ones they will have their entire life. Attempts to spare vessels are of utmost importance and the preservation of already placed catheters requires careful review prior to removal. Chronic and complex medical conditions often require a child to rely on a vascular access device for extended periods of time. Because vessels frequently become impaired and unavailable for future use, maintaining the quality and functionality of venous access can be a challenge.

One of the first indications of a potential complication with a vascular access device is a line that does not draw back blood appropriately or when a saline flush

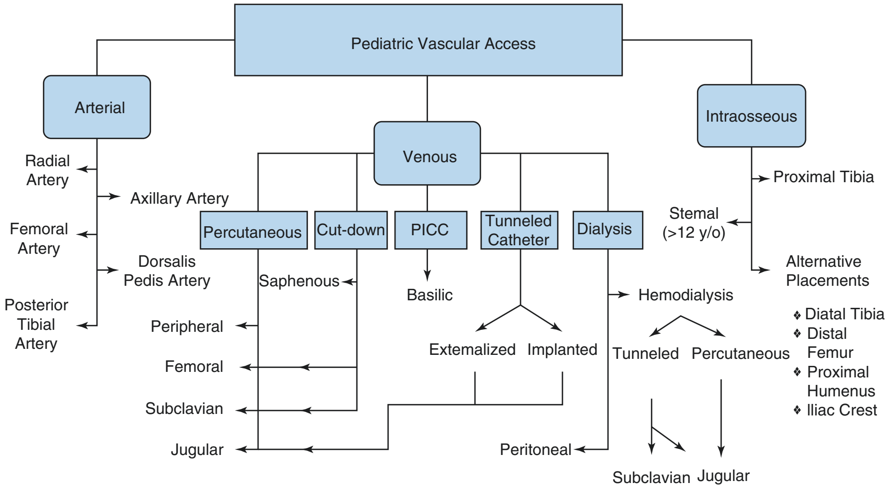

Pediatric Vascular Access
Arterial
Radial Artery
Axillary Artery
Femoral Artery
Dorsalis Pedis Artery
Posterior Tibial Artery
Venous
Percutaneous
Cut-down
PICC
Tunneled Catheter
Dialysis
Saphenous
Basilic
Peripheral
Femoral
Subclavian
Jugular
Extemalized
Implanted
Hemodialysis
Tunneled
Percutaneous
Peritoneal
Subclavian
Jugular
Intraosseous
Proximal Tibia
Stemal (>12 y/o)
Alternative Placements
❖ Diatal Tibia
❖ Distal Femur
❖ Proximal Humenus
❖ Iliac Crest

**Fig. 4** Vascular Access Overview in Children (Kody L Moore and Christopher P Coppola)

or medication administration attempt is difficult or unsuccessful. An X-ray can be performed to evaluate for migration or kinking of the line. When migration is not found, an occlusion must be considered. If identified early, flushing the line, sometimes with a heparinized saline solution, may resolve the occlusion. Treatment with tissue plasminogen activator, a serine protease, may possibly resolve thrombosis. Repositioning the patient, especially into Trendelenburg position may give a spontaneous return of blood flow and improve flushing. The use of soft guide wires/dilators, a balloon, and aspiration thrombectomy have been performed with success in salvaging venous catheters that have developed fibrin sheaths in children. Consultation with interventional radiology can be beneficial in these situations. Attempts at resolving the obstruction and establishing patency should be performed prior to replacement of a catheter in an uninjured vein.

Catheter-related infections (CRI) are of importance not only to the preservation of the line but to the health risk associated with these infections. Removing the source of infection is vital to treatment; however, this can create a conflict for those patients that are catheter dependent for chronic therapy. Bacteria can enter a line by several methods including the insertion point and the hub of the catheter during each attempted access. Limiting the number of accesses to the line, having staff familiar with line care, and proper sterilization of the hub can decrease the risk of line contamination. Antibiotics during initial placement do not decrease the risk of development of CRI. However, when infection is suspected, broad-spectrum antibiotics should be initiated to preserve the line. Removal of the line may become necessary if fever continues more than 48 h after starting antibiotics. Blood cultures of both central and peripheral catheters are crucial to the diagnostic process. There has been development of antibiotic-impregnated catheters which can be used during initial insertion. Using these catheters has shown a reduction in CRI related removals. Children who are immunocompromised, have known infectious processes, or have had previous premature catheter removals due to CRI could benefit with the insertion of these types of lines. Antibiotic impregnated catheters may also be considered when over-wiring replacements of infected catheters are attempted. The use of antibiotic and ethanol locks have also been used to salvage infected lines as well as prevent CRI related infections.

Mechanical issues can arise in any line placement. Dislodgement is a common reason for mechanical failure, especially in young, active children. Even cuffed lines are not immune to accidental removal. Many dislodgements can be avoided with proper securing of the line with an adhesive device or suture, as well as protecting the exterior portion of the line with dressings or a garment. Exchange over a guide wire can be beneficial for kinked catheters and those found to be leaking, thus allowing for replacement of the catheter while preserving the accessed vein. When a child is known to have a chronic disease in which there is high likelihood for repeated need for vascular access points, a venous access plan can be created to help guide future considerations for removal and insertion of any catheter. The use of imaging can help create a baseline of patent and usable vessels. One should consider obtaining this early, before multiple accesses are performed.

## Study Questions

Question 1) When placing an intraosseous (IO) access device, which of the following is not a contraindication to placement?

A. Overlying Cellulitis
B. Overlying Tattoo
C. Fracture of entry site bone
D. Recent prior IO attempt at entry bone

Answer: B) Overlying Tattoo. When placing an IO access device, any infection including the skin and soft tissue would exclude the location as an entry point. A fracture or prior recent attempts of IO placement at an entry bone are also contraindications. A patient tattoo of the overlying skin does not prevent placement of an IO.

Question 2: When placing a radial arterial line, which of the following tests should be performed to ensure adequate dual arterial flow is present prior to placement?

A. Buerger Test
B. Hess Test
C. Romberg Test
D. Allen Test

Answer: D) Allen Test. Prior to attempted placement of a radial arterial line, both radial and ulnar arteries should be compressed, hand squeezed to blanch skin, and ulnar artery released. Known as the Allen test, proper dual arterial flow is noted when there is color return to the hand within 5 seconds of ulnar artery releasement. Abnormal tests require consideration for alternative placement.

Question 3: After venotomy is performed and the central catheter is placed, radiologic imaging is used to confirm proper placement. Which of the following locations is consistent with a properly placed catheter tip?

A. Pericardial Sac
B. Internal Jugular
C. Superior Vena Cava
D. Right internal mammary

Answer: C) Superior Vena Cava. With placement of a central catheter, it is important to use imaging studies to ensure the catheter tip is at an appropriate location to reduce the risk of complications. The optimal location of a central venous catheter tip is located at the cavo-atrial junction with acceptable locations including Superior vena cava, right atrium, and inferior vena cava.

## Further Reading

Ares G, Hunter CJ. Central venous access in children: indications, devices, and risks. Curr Opin Pediatr. 2017;29(3):340–6. https://doi.org/10.1097/mop.0000000000000485.

Baskin KM, Mermel LA, Saad TF, et al. Evidence-based strategies and recommendations for preservation of central venous access in children. J Parenter Enter Nutr. 2019;43(5):591–614. https://doi.org/10.1002/jpen.1591.

Church J, Jarboe M. Vascular access in the pediatric population. Surg Clin North Am. 2017;97(1):113–28.

Farrelly JS, Stitelman DH. Complications in pediatric enteral and vascular access. Semin Pediatr Surg. 2016;25(6):371–9.

Gupta N, Gandhi D, Sharma S, Goyal P, Choudhary G, Li S. Tunneled and routine peripherally inserted central catheters placement in adult and pediatric population: review, technical feasibility, and troubleshooting. Quant Imaging Med Surg. 2021;11(4):1619–27. https://doi.org/10.21037/qims-20-694.

Hahn RG, Prough DS, Svensen CH. Perioperative fluid therapy. 1st ed. Hoboken, NJ: CRC Press; 2006.

Jöhr M, Berger TM. Venous access in children: state of the art. Curr Opin Anaesthesiol. 2015;28(3):314–20. https://doi.org/10.1097/ACO.0000000000000181.

Lyon S, Given M, Marshall N. Interventional radiology in the provision and maintenance of long-term central venous access. J Med Imaging Radiat Oncol. 2008;52(1):10–7. https://doi.org/10.1111/j.1440-1673.2007.01904.x.

Naik V, Mantha SSP, Rayani B. Vascular access in children. Indian J Anaesth. 2019;63(9):737–45. https://doi.org/10.4103/ija.ija_489_19.

# Gastrostomy and Enteral Access

**Kelsey R. Tieken, Katharine A. Armstrong, Meghana V. Kashyap, and Angela M. Hanna**

**Key Points**

1. Patients under consideration for long-term enteral access require thorough nutritional assessment. When possible, maximize oral nutrition prior to surgical intervention.
2. Multiple types of enteral access exist and the decision regarding which type to utilize depends on patient factors and the duration of anticipated need for enteral access. Patients requiring greater than 3 months of enteral support should be evaluated for surgical gastrostomy tube placement. Gastrostomy tubes may be placed with the aid of endoscopy: percutaneous endoscopic gastrostomy (PEG) or surgically either open or laparoscopically. The appropriate approach depends heavily on patient factors.
3. Other, less commonly utilized types of enteral access exist and are indicated in patients with specific pathologies. For example, patients with high aspiration risk or severe GERD may benefit from post-pyloric feeding.
4. Timing of use of tubes after placement vary based on surgeon preference. Caregiver education regarding routine maintenance and use of tubes is essential.

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5. Complications of tube placement are generally minor (skin break down, tube clogging), etc.) and can be managed with supportive care and proper tube maintenance. Major complications (tube dislodgement) are rare but can become a surgical emergency.
6. Timing and setting of tube removal and exchange vary depending on the type of tube in place. Surgical intervention may be necessary if tubes are still leaking 4 weeks post-removal.

1. Background and Etiology of Malnutrition
    a. Pediatric patients who are ill or have inadequate nutrition are usually first offered dietary adjustments for oral feeds. However, patients who have underlying structural or physiologic abnormalities that preclude oral feeding should be considered for early enteral nutrition. There are multiple options for enteral access, most commonly used is the gastrostomy tube.
    b. When working up a patient for potential long-term enteral access, one must begin with a thorough nutritional assessment based on the patient's underlying diagnosis and expected hospital course.
        i. Long-term enteral access should be considered in patients with impaired swallowing or oral motor development often seen with neuromuscular impairment (e.g., cerebral palsy), prematurity, anoxic brain injury, seizure disorders, and some chromosomal abnormalities.
        ii. Patients with impaired absorption or digestion such as short bowel syndrome, IBD, biliary atresia, or inborn errors of metabolism tend to have higher metabolic needs, as well as patients with congenital heart disease or cystic fibrosis.
        iii. It may also be considered for patients who cannot sustain adequate oral intake to meet their higher metabolic demands. Pediatric patients with sepsis, critical illness, post-operative states, burns, or cancer benefit from enteral nutrition and may additionally have other underlying comorbidities that prevent oral intake.
2. Diagnosis of Malnutrition:
    a. Assessment of a patient's current nutritional status is also a key component of workup for enteral access. It is important to assess where the patient falls on the pediatric growth curve and how they are tracking for their expected growth.
        i. A multidisciplinary approach should be utilized with the early involvement of registered dieticians to evaluate patients who are at significant risk for malnourishment and caloric intake carefully tracked.
        ii. Options to improve caloric intake include offering higher-calorie foods, adding oral supplements, or boosting nutritional density of foods by adding macronutrients to meals.

    iii. If the patient is unable to take nutrition orally adequately or safely, consideration must be given to orogastric (OG) or nasogastric (NG) tubes, endoscopically or surgically placed gastrostomy tubes, or gastrojejunostomy tubes.

3. Management of malnutrition: Establishing Appropriate Enteral Access

    a. When determining which type of enteral access a patient may need, consider the anticipated duration of support needed.
    b. Patients who require short-term enteral access (less than three months) should be considered for orogastric (OG) or nasogastric (NG) tube placement.

        i. OG tubes are often used in intubated patients and are placed through the mouth and advanced into the stomach.
        ii. NG tubes are placed through the nasal passage and advanced into the stomach and may be bridled around the nasal septum in young children to prevent from being inadvertently dislodged or pulled out.
        iii. It is important to confirm positioning below the gastroesophageal junction with abdominal X-ray after both OG and NG placement.

    c. For patients who require long-term enteral access (greater than three months), surgical enteral access should be considered. If the patient is not already receiving tube feeds, an NG tube may be placed for a few days minimum to ensure tolerance of gastric feeds.

        i. When assessing the patient for surgery, an upper GI study should be obtained to identify the anatomy, evaluate for hiatal hernia, confirm the location of ligament of Treitz and rule out congenital malrotation to help aid surgical planning.

    d. Several options exist for surgical enteral access. Gastrostomy tubes are commonly utilized in the pediatric population.

        i. Anatomy: Gastrostomy tubes are placed directly through rectus midway between umbilicus and costal margin and enter the antrum of stomach away from pylorus and GE junction
        ii. The most commonly used techniques for gastrostomy tube placement are percutaneous endoscopic gastrostomy (PEG), open gastrostomy, and laparoscopic gastrostomy.

    e. Gastrojejunostomy (GJ) tubes may be used when gastric feeds are not tolerated or in cases of severe reflux. However, GJ tubes are more prone to dislodgement and require frequent replacement (interventional radiology (IR), endoscopically, open). Jejunostomy tubes may also be considered but are less commonly placed in pediatrics due to the risk of volvulus around the single lead point.

4. PEG Tubes
    a. Percutaneous endoscopic gastrostomy tube placement is a commonly utilized technique for enteral access in children.
    b. Benefits: minimally invasive technique, short procedure time, relatively decreased cost, and a shorter recovery period.
    c. Risks: relatively blind nature of the procedure, increased risk of early dislodgement and gastric spillage, and inability to complete the procedure safely requiring conversion to laparoscopic-assisted or open placement of gastrostomy tube.
    d. Contraindications: history of prior abdominal surgery (relative), congenital malrotation or other foregut anatomical variances (e.g., esophageal atresia), inability to tolerate gastric feeds, or need for concomitant abdominal procedures at the time of placement.
    e. The technique for PEG tube placement is described below:
        i. Anesthesia is induced and a pediatric upper endoscope is placed. Perform a complete upper endoscopy to ensure there are no anatomic abnormalities not previously recognized.
        ii. Maximally insufflate the stomach to bring it directly against the abdominal wall and face the lighted scope toward the desired area of tube placement. A second surgeon looks for an appropriate area for placement by looking for transillumination of the scope through the abdominal wall. The site is confirmed with palpation of the abdominal wall over the area of transillumination and visualizing indentation of the gastric wall with the endoscope.
        iii. Local anesthetic is injected into the area, a small skin incision is made over the site and the stomach is accessed under direct visualization with the endoscope with a large bore needle. A snare is passed through the endoscope and a guidewire is placed through the needle into the stomach. The wire is grasped with the snare and pulled out through the esophagus and mouth by the endoscopist.
        iv. The wire is secured to the PEG device and the device pulled back through the esophagus into the stomach and through the gastric wall into position.
        v. The internal bolster should rest securely against the gastric wall. An external bumper is then placed and secured into position on the abdomen with or without non-absorbable sutures. The endoscope should again be introduced to confirm the proper positioning of the tube (Fig. 1).

5. Surgical Gastrostomy Tubes
    a. Laparoscopic gastrostomy tube placement is a common procedure in pediatric surgery.
    b. Benefits: direct visualization, decreased risk of tube dislodgement through gastropexy, and the ability to perform multiple procedures at the same time. For example, many patients who require enteral access also suffer from

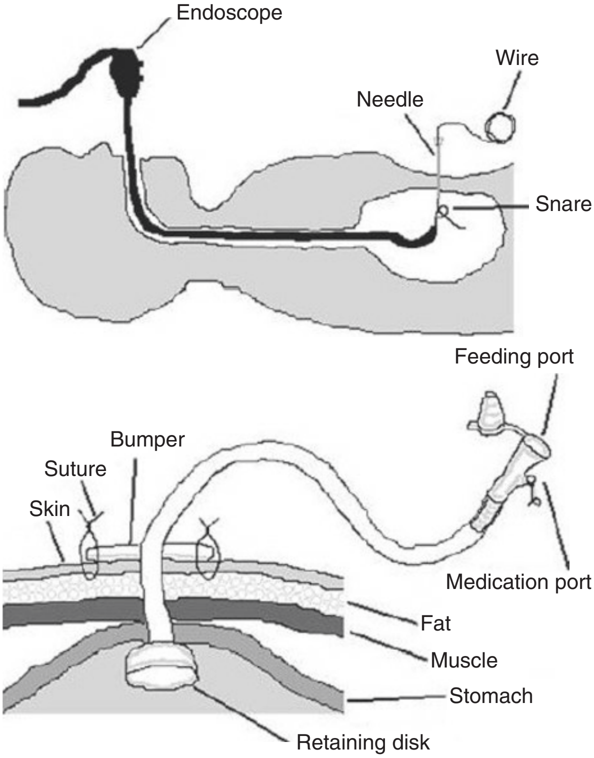

**Fig. 1** Technique of percutaneous endoscopic gastrostomy placement. Source: Bryan Walters and Christopher Coppola

severe reflux, and a laparoscopic fundoplication may be performed at the same time in these patients.

c. Risks: need for general anesthetic, relatively increased cost, creation of multiple incisions, longer recovery time, and the potential for conversion to open procedure.
d. Contraindications: prior abdominal surgeries with high risk for significant adhesions (relative), inability to tolerate general anesthetic, and inability to tolerate gastric feeds (relative).
e. One technique for laparoscopic gastrostomy tube placement is described below; however, technique may vary between different institutions or based on surgeon preference:

   i. General anesthesia is initiated. The abdomen is accessed via a periumbilical incision and pneumoperitoneum is established. The laparoscope is inserted, and the abdomen inspected.
   ii. A full-thickness stab incision is made in the LUQ where the gastrostomy placement is planned. An atraumatic grasper is placed through the incision and the greater curvature of the stomach grabbed and elevated toward the abdominal wall, taking care to stay well away from the pylorus.

    iii. Two trans-fascial sutures are placed through the abdominal, through the anterior wall of the stomach, and back out the anterior abdominal wall on either side of the blunt grasper and the stomach is released.
    iv. A large bore needle is placed through the gastrostomy tube site between the two sutures and into the anterior wall of the stomach. A wire is advanced through the needle, an appropriately sized tract is created with serial dilations and the gastrostomy tube is then placed over the wire using the Seldinger technique. The balloon is adequately inflated, and visualization is confirmed within the stomach.
    v. The trans-fascial sutures are then secured to pexy the stomach to the abdominal wall. (Most surgeons then cut these sutures within 3–5 days after placement.) The laparoscope is removed, and fascia and skin are closed (Fig. 2).

f. A separate technique for laparoscopic gastrostomy tube placement involves bringing a portion of the stomach out through a trocar site.

    i. The laparoscopic is inserted through the umbilical trocar as described above, creating a trocar site in the LUQ where the tube will be placed, then grasping the gastric wall and bringing it directly out through the trocar site. A purse string suture is then placed around the gastrostomy tube insertion site in the stomach, and then the stomach is tacked in four quadrants to the underside of the fascia, and the stomach is returned to the abdominal cavity.
    ii. A gastrotomy is then created in the center of the purse string and the tube is placed through the hole. If either the fascia or skin opening around the tube is larger than the tube itself, it must be closed around the tube to prevent leakage.

g. Open gastrostomy tube placement is another option for gastrostomy tube placement.

    i. Benefits: ability for direct visualization of the tube entering the gastric lumen, allows for a smaller tube insertion site which then decreases the risk of leakage around the tube, and the gastric wall can be pexied to the anterior abdominal wall to help prevent dislodgement of the tube. In addition, if other open procedures are indicated at the time of the tube placement, they can be performed at the same time and minimize multiple trips to the operating room.
    ii. Risks: risk of general anesthetic, increased cost compared to more minimally invasive surgical approaches, and increased pain and longer recovery time with the larger incision.
    iii. Contraindications: inability to tolerate gastric feeds and inability to tolerate a general anesthetic (relative).

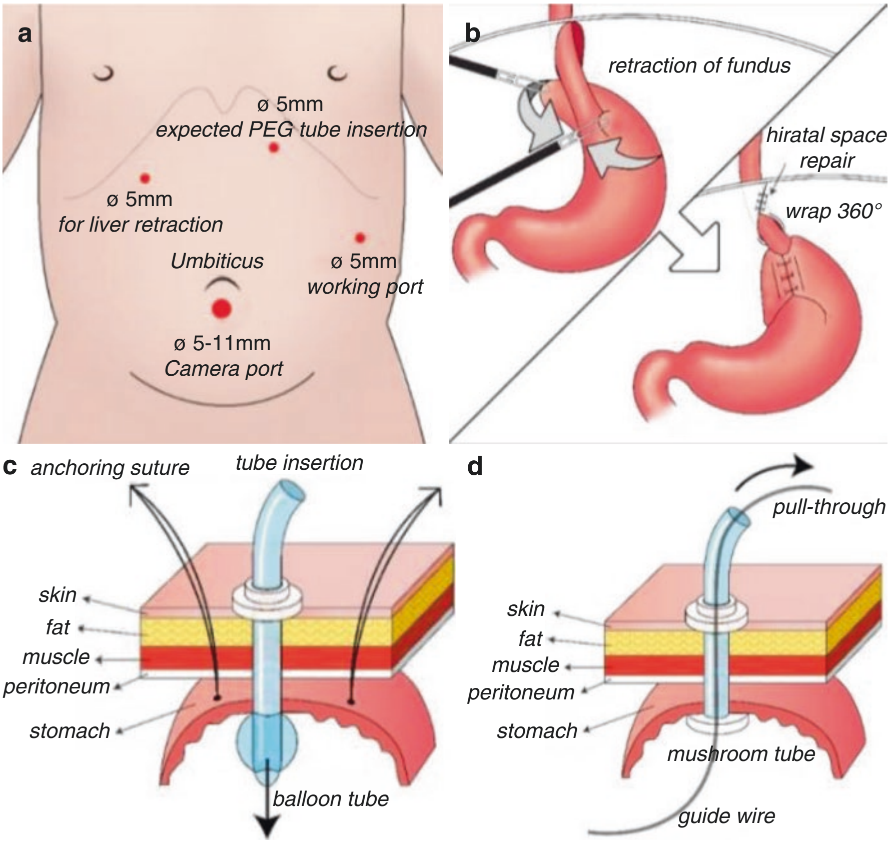
a
ø 5mm
expected PEG tube insertion
ø 5mm
for liver retraction
Umbiticus
ø 5mm
working port
ø 5-11mm
Camera port
b
retraction of fundus
hiratal space repair
wrap 360°
c
anchoring suture
tube insertion
skin
fat
muscle
peritoneum
stomach
balloon tube
d
pull-through
skin
fat
muscle
peritoneum
stomach
mushroom tube
guide wire

**Fig. 2** Technique of laparoscopic gastrostomy tube placement with concomitant gastric fundoplication. (**a**). Location of laparoscopic ports (**b**). Laparoscopic gastric fundoplication (**c**). Transfascial sutures secure the stomach to the anterior abdominal wall so that the gastrostomy tube passed through the left upper quadrant port site can be advanced through a gastrotomy to the gastric lumen. (**d**). The gastrostomy tube can also be placed in a technique similar to percutaneous endoscopic gastrostomy where the tube is pulled through the abdominal wall over a guide wire placed through a needle. Source: Yoon S, Kim S-H, Lee YJ, Park JH, Cho YH, Chung JH. Experience with a Hybrid Procedure Involving Laparoscopic Fundoplication with Percutaneous Endoscopic Gastrostomy in Chronically Ill Children. J. Clin. Med. 10(19):4291, 2021. Used under Creative Commons Attribution 4.0 International license (CC BY 4.0) https://creativecommons.org/licenses/by/4.0/ with adaptation of figure legend

iv. The technique for placement of the open gastrostomy tube is essentially the same as it is for laparoscopic, except that a laparotomy incision must be made rather than multiple small incisions for laparoscopic trocar placement. Care must be taken to appropriately position the incision so that once closed, it allows for adequate healing of both surgical sites.

6. Other procedures
    a. Although the PEG tube and gastrostomy tubes are the most common methods for obtaining enteral access, other types of interventions also exist for special populations.
        i. In a patient with severe reflux or who is a high aspiration risk, postpyloric feeding may be indicated. Oroduodenal or nasoduodenal tubes may be used for short-term access.
        ii. Gastrojejunostomy tubes are also an option. These are similar to gastrostomy tubes, but the catheter is extended beyond the internal balloon into the proximal small intestine. Unfortunately, these frequently become dislodged or move out of correct positioning, and therefore often require frequent replacements.
        iii. Use of jejunostomy tubes is uncommon as they can cause volvulus around the tube which acts as a lead point.
    b. For certain patients, gastric bypass, Roux-en-Y bypass with gastrostomy tube placement, or esophagojejunostomy procedures may be indicated.
7. Complications
    a. Many minor common complications from gastrostomy tube placement exist; most are related to issues with skin irritation or breakdown or the tube itself.
    b. Granulation tissue can develop around the insertion site of the tube which can cause bleeding or leaking around the tube.
        i. In general, small areas of granulation tissue can be cauterized with silver nitrate. Triamcinolone cream can be applied up to three times per day, alternating one week on and one week off for two weeks of total treatment.
        ii. Severe cases, or cases that fail initial management may require surgical debridement. In these cases, it is important to avoid debriding enough tissue to significantly increase the size of the insertion site, as this will lead to leakage of gastric contents around the tube.
    c. Skin irritation is common around tubes, especially if the external bumper is incorrectly placed. A clinical exam is important to rule out cellulitis or an underlying abscess around the site.
        i. Treatment involves keeping the underlying skin clean and dry and the tube perpendicular to the skin. In general, this will resolve with local wound care and the use of protective barriers to prevent further irritation during the healing process.
        ii. If cellulitis or an abscess is present, antibiotic treatment should be initiated in addition to local wound cares. If an abscess is suspected based on clinical exam, follow-up imaging and incision and drainage may be required.

d. Pyloric obstruction is uncommon in a correctly placed tube, but if present can be managed initially by partially deflating the balloon.
   i. If persistent, surgical revision of the gastrostomy tube site is indicated.

e. Complications of the tube itself are also common. One of the most commonly encountered problems is tube leakage.
   i. If a tube is noted to be leaking, first check to ensure that it is not clogged and then obtain a tube study to ensure it is appropriately placed.
   ii. Leakage may also be caused by pressure from the internal bumper on the abdominal wall if the stem of the tube is too short. If this is the case, the tube may need to be exchanged or the external bumper adjusted depending on the type of tube. Ensure that the external bumper is appropriately flush with the skin and perpendicular to the abdominal wall.
   iii. In cases of tube leakage, never increase the size of the tube. If the insertion site is too large, remove the tube for a tube holiday and allow time for the site to shrink.

f. Tube clogging is another common problem with gastrostomy tubes.
   i. If the tube is clogged, first flush with 60mL of warm water. If unsuccessful, Coca-Cola or pancreatic enzymes instilled in a bicarbonate solution can be flushed into the tube and then flushed with warm water again two to three minutes later.

g. Dislodged tubes are another commonly encountered problem and require immediate replacement to prevent closure of the tract.
   i. If a tube is dislodged within the first few weeks of placement (and thus the tract is immature), it is a surgical emergency and should be treated as a perforated viscous.
   ii. In certain circumstances, it may be possible to try to attempt replacement followed by a radiographic tube check to ensure it is correctly placed into the stomach.
   iii. Replacement may also be attempted via endoscopy to find the hole and replace via a similar method as PEG tube placement.
   iv. However, if these options fail or if the patient becomes unstable, this should also be treated as a perforated viscous as the stomach may have fallen away from the abdominal wall and gastric contents can spill into the peritoneal cavity.

h. Over time, it is common that a tube will start to deteriorate, as evidenced by discoloration, irregular beading of the tube or a malodorous smell. Eventually, this can cause leaks or breaks in the tube. It is recommended that tubes be replaced once any dysfunction begins to affect the administration of required medications or enteral nutrition.

i. Major complications: Major complications of gastrostomy tube placement are rare, but important to understand and take into consideration if a patient presents with an unexpected post-operative course.
    i. When placing PEG tubes, one serious complication includes placing the tube directly into the colon rather than the stomach or placing the tube through the colon into the stomach. Care must be taken to ensure strong transillumination against the abdominal wall and direct one-to-one visualization of the gastric wall indentation while palpating from the abdominal wall.
    ii. Other major complications that may arise from any type of tube placement include gastroenteric fistula, peritonitis, either from a missed enterotomy intra-operatively or early tube dislodgement, severe bleeding, and benign pneumoperitoneum.

## Additional Notes

Tube use and Maintenance: Regimens regarding tube use for medication administration and enteral nutrition vary based on surgeon preference. In general, it is felt to be safe to use PEG tubes immediately after placement. Surgically placed gastrostomy tubes usually institute a waiting period of hours or days prior to initiation of feeds. Once deemed appropriate, feeds are slowly initiated and slowly increased to goal as tolerated.

Tube changes are generally completed once the tract has fully matured, which is approximately six to eight weeks following initial placement. The first tube change must be done by a physician; however, subsequent changes may be done by a properly educated caregiver or pediatrician every three months. It is important to note that for tubes that have an internal bumper that is a balloon, the balloon must be inflated with water. Saline can degrade the balloon faster than water causing tube malfunction and should never be used to insufflate. Caregivers must also be thoroughly educated on routine cares of the tube, including monitoring for proper bolster placement to prevent the breakdown of the skin and regular cleaning of the gastrostomy tube site. Frequent flushing is also important to prevent clogging of the tube. Bulky agents or resins should not be administered via the tube as they will cause it to clog. Medications given through the tube must be in solution form or crushed and dissolved in water. The tube should be flushed with 20 mL of water after administration of any medication or formula.

Tube removal/exchange: Tubes older than two months often can be replaced in an office setting. If a tube replacement at any time is difficult or uncertain, a contrast study should confirm the tube position. When a tube needs to be exchanged, it is important to know what type of tube the patient has. Review operative reports if they are available, but if the tube type is unknown, it may be necessary to perform an endoscopy prior to removal to determine which type of internal bumper is present to ensure safe removal. In pediatrics, most internal bumpers are balloons; however, multiple other types do exist. If a balloon is present, in order to remove the existing tube, simply deflate the balloon entirely and remove the tube. A new tube may be placed directly through the existing tract and the external bumper secured at the proper height.

When a patient no longer requires their tube, it is generally safe to remove them in an office setting unless the tube in place is a PEG tube. A PEG tube that must be removed or replaced may require endoscopy. After tube removal, apply a barrier cream to the skin around the insertion site and cover with gauze. There may be some drainage from the site initially. If this does not resolve by four weeks, then surgical closure of the site is usually necessary.

## Study Questions

1. An 8-week-old ex-30-week premature infant is in the NICU. She has a history of CHD and FTT. She is currently tolerating tube feeds via NG at goal rate. You have been consulted for evaluation for long-term enteral access. What is your next step in the management of this patient?

   a. Obtain UGI study
   b. Placement of PEG tube
   c. Perform esophagogastroduodenoscopy
   d. Place a surgical gastrostomy tube

   Answer: (a.) Obtain UGI

   1. When working up a patient for gastrostomy tube placement, several considerations must be made. First, it is important to ensure the patient will be able to tolerate tube feeds. This patient is already tolerating tube feeds at goal rate via NG tube. Second, it is important to assess for anatomic abnormalities that may affect surgical options for enteral access. This can be readily accomplished via UGI study.

2. A 2-year-old male is brought to the ER by his mother for “feeding tube issues.” He has a history of cystic fibrosis and had a laparoscopic gastrostomy tube placed as an infant due to malnutrition with the inability to sustain adequate oral intake. He is currently receiving nutrition through a combination of oral feeds during the day and tube feeds at night. His mom states she tried to run his tube feeds earlier this evening, however, nothing would go through. She tried to flush the tube with water but was unsuccessful. What is the next step in management?

   a. Remove the tube
   b. Attempt to flush again with water
   c. Flush tube with pancreatic enzymes instilled in bicarbonate solution
   d. Consult IR for emergent tube exchange

Answer: (c.) Flush tube with pancreatic enzymes instilled in bicarbonate solution

1. Tube clogging is a common issue for patients who are receiving long-term enteral access via gastrostomy tube. If the tube is unable to be flushed with warm saline, the next step is to flush the tubing with pancreatic enzymes instilled in bicarbonate solution. It is important to instruct caregivers on proper tube maintenance such as flushing the tubing after administration of meds or feeds to prevent this complication; however, this can happen even with proper tube maintenance.

3. You are called to the bedside of a patient who is POD4 from a laparoscopic gastrostomy tube placement. The surgery was uneventful, and the patient had been tolerating tube feeds. When the nurse came into the room at the beginning of the shift, he found the gastrostomy tube laying at the patient's side. It is unclear how long the tube was out. When you arrive at the patient's room you indeed find the tube has become completely dislodged. What is your next step in management?

   a. Place a catheter into the tract to keep it patent
   b. Obtain a contrasted study to evaluate for extravasation into the peritoneum
   c. Immediately replace a tube at bedside and obtain a tube study
   d. Return to the OR

   Answer: (d.) Return to the OR

   1. This patient had a laparoscopic gastrostomy tube placed within the last week, and the tube has now become completely dislodged. This should be treated as a perforated viscous as there has not been enough time for a tract to form between the stomach and the abdominal wall, and there is a significant risk of gastric spillage if the stomach pulls away from the abdominal wall warranting swift return to OR.

## Further Reading

Axelrod D, Kazmerski K, Iyer K. Pediatric enteral nutrition. J Parenter Enter Nutr. 2006;30:S21–6.

Beres A, Bratu I, Laberge J. Attention to small details: big deal for gastrostomies. Semin Pediatr Surg. 2009;18(2):87–92.

Chung DH, Georgeson KE. Fundoplication and gastrostomy. Semin Pediatr Surg. 1998;7(4):213–9.

Enteral Access | PedSurg Resource. https://www.pedsurglibrary.com/apsa/view/PedSurg%20Resource/1884018/all/Enteral_Access (Downloaded 5 July 2021).

Gastrostomy tubes: Placement and routine care | UpToDate Resource. https://www.uptodate.com/contents/gastrostomy-tubes-placement-and-routine-care (Downloaded 9 July 2021).

Overview of enteral nutrition in infants and children | UpToDate Resource. https://www.uptodate.com/contents/overview-of-enteral-nutrition-in-infants-and-children (Downloaded 9 July 2021).

Vervloessem D, van Leersum F, Boer D, Hop W, Escher J, Madern G, Bax K. Percutaneous endoscopic gastrostomy (PEG) in children is not a minor procedure: risk factors for major complications. Semin Pediatr Surg. 2009;18(2):93–7.

# Chest Tube

**Luiz G. Foernges**

**Key Points**

1) A chest tube or tube thoracostomy is used to drain air, blood, or fluid in the thoracic cavity between the visceral pleura of the lung and the parietal pleura of the chest wall.
2) In emergent conditions, such as tension pneumothorax, clinical signs of lung collapse are enough indication to place a needle decompression of the tension pneumothorax followed by a chest tube.
3) While placing a chest tube, care must be taken to follow the anatomic landmarks of the chest to avoid injuring thoracic or abdominal organs.

Tube thoracostomy is indicated for drainage of air, blood, or fluid accumulated between visceral and parietal pleura.

1. Indications:
    - (a) Emergent:
        - (i) Tension pneumothorax
    - (b) Urgent:
        - (i) Pneumothorax in patients on mechanical ventilation
        - (ii) Tension pneumothorax after needle decompression

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        (iii) Pleural effusions affecting respiratory function
        (iv) Traumatic hemopneumothorax

    (c) Elective:

        (i) Empyema or complicated parapneumonic pleural effusion
        (ii) Malignant pleural effusion
        (iii) Transudates from cardiac, renal, and hepatic diseases
        (iv) Postoperative after cardiac and thoracic procedures

2. Contraindications:
    (a) Coagulopathy
    (b) Pulmonary bullae
    (c) Pulmonary collapse “whiteout” from mucus plug
    (d) Dense pleural adhesions
    (e) Drainage of a post-pneumonectomy space

3. Diagnosis:
    (a) Clinical exam: primary diagnosis for tension pneumothorax. Absent breath sounds, deviation of the trachea away from the side with the tension pneumothorax, hyper-expansion of the chest side with tension pneumothorax, increased venous pressure with dilated neck veins, and cardiorespiratory collapse.
    (b) Chest X-ray: initial diagnostic test, cheap, fast, and readily available. Will require lateral and/or decubitus views for small effusions. Bilateral effusions are more commonly associated with transudates. Can also suggest etiology of the effusions (consolidation, tumors, and cardiomegaly).
    (c) Ultrasonography: available in most emergency departments and intensive care units. Operator dependent. Able to detect small amounts of pleural fluid. Useful tool for multiloculated pleural effusions and identifying landmarks before chest tube placement, especially in small children.
    (d) Computed tomography: more sensitive to differentiate pleural thickening and focal pleural masses. Main disadvantage is radiation exposure.
    (e) Magnetic resonance imaging: limited role on diagnosis of pleural disease.
    (f) Pleural fluid analysis: pleural fluid can be obtained prior to chest tube placement to help diagnose and guide treatment. Pleural fluid samples can be sent for protein, lactate dehydrogenase, glucose, pH, amylase, hematocrit, cell count and differential, triglycerides, cholesterol, chylomicrons, culture, Gram stain, and cytology analysis.

4. Equipment:
    (a) Mask, head cover, sterile gloves, and gown
    (b) Skin antiseptic solution
    (c) Sterile drapes
    (d) Gauze
    (e) Syringes and needles

  (f) Scalpel and blade
  (g) Suture:
    (i) Preterm and term infants #3-0 non-absorbable suture
    (ii) 1–6 years #2-0 non-absorbable suture
    (iii) Above 6 years #0 non-absorbable sutures
  (h) Instruments for blunt dissection, curved hemostatic and Kelly clamps
  (i) Needle driver
  (j) Mayo scissors
  (k) Chest tube:
    (i) Preterm infants: 8Fr
    (ii) Term infants: 10Fr
    (iii) 1–3 years of age: 12Fr
    (iv) 3–10 years of age: 16–18Fr
    (v) 10–15 years of age: 20–28Fr
    (vi) Above 15 years of age: 28Fr
  (l) Connecting tube
  (m) Closed drainage system
  (n) Adhesive tape

5. Procedure:
  (a) Review imaging. Check and confirm the side of the procedure.
  (b) Administer intravenous analgesia and sedation. Prophylactic antibiotics should be given in trauma cases.
  (c) Position patient supine, with the arm on the side of the procedure abducted 90° with the palm of the hand resting under or near the head, secure the wrist with adhesive tape or soft restraint, being sure that the hand is perfused.
  (d) Identify the landmarks, "safe triangle":
    (i) Inferior: level of nipple (4th intercostal space), placement of chest tube below the 4th intercostal space will increase the chance of intra-abdominal chest tube placement.
    (ii) Anterior: lateral border of pectoralis major muscle, placement of chest tube anterior to this line will increase the chance of injury to the pectoralis muscle and breast tissue.
    (iii) Posterior: anterior border of the latissimus dorsi, placement posterior to this line will increase the chance of injury to the long thoracic nerve.
  (e) Ultrasonography can guide insertion site. Select a place within the safe triangle, close to the midaxillary line.
  (f) Use 1 % lidocaine (maximum dose 3.3–4.4 mg/kg); infiltrate a wheal for the skin. Redirect the needle to the expected course of the chest tube toward the superior border of the rib injecting subcutaneous tissue, intercostal muscle, and pleura.

- (g) After entering the pleura with the needle, aspirate air, blood or pleural effusion to confirm position, if necessary, aspirate fluid for diagnosis after anesthesia.
- (h) Make a small incision following the curvature of the rib (approximately the diameter of the chest tube) in the skin overlying the rib that is below the intercostal space to be used for placement of the chest tube.
- (i) Bluntly dissect the subcutaneous tissue with a hemostatic clamp, spreading the tissue. Direct the tip of the hemostat toward the superior border of the rib. Palpate the tract with a finger to be sure of the correct intercostal space, and superior border of the rib during dissection. Small bore chest tubes can also be inserted using Seldinger technique over guidewire and dilators.
- (j) Gently advance the hemostat over the superior border of the rib, opening the hemostat in the same direction of the ribs to dissect the intercostal muscles.
- (k) With the hemostat closed, carefully pierce the pleura, and spread in a controlled manner not to injury intrathoracic organs. A rush of air or fluid may occur when the pleura is entered.
- (l) For older children, palpate the tract with a finger, passing the finger through the intercostal space into the pleural cavity to evaluate the presence of adhesions.
- (m) Measure the length between the insertion site and the apex of the lung to estimate the length of the chest tube, place a clamp on the chest tube to mark the estimated length measured.
- (n) Grasp the fenestrated portion of the chest tube with a Kelly clamp and introduce it in the pleural space, release the Kelly clamp and gently advance the chest tube posteriorly and superiorly. Be sure that all the side holes in the chest tube are within the pleural cavity.
- (o) Holding the tube in place, connect the chest tube to the drainage device. Release the clamp from the chest tube. Connect the closed drainage system to suction, if indicated.
- (p) Secure the chest tube to the skin with non-absorbable sutures. Approximate the skin around the chest tube with sutures. Place sterile dressing around the chest tube site. Strap the chest tube to the flank with adhesive tape to prevent kinking and tension. Tape chest tube connections to prevent dislodgement.
- (q) Obtain chest radiography to ensure correct placement.

6. Complications:

   - (a) Severe:

     - (i) Puncture of the heart, lung, liver, and spleen
     - (ii) Damage to intercostal vessels, anterior thoracic artery, and trachea
     - (iii) Wrong side

- (b) Moderate:
  - (i) Infection
  - (ii) Nerve damage
  - (iii) Chest tube disconnection or displacement
- (c) Mild:
  - (i) Subcutaneous emphysema
  - (ii) Poor drain management
  - (iii) Underwater seal not maintained

7. Chest tube removal:
   - (a) Remove dressings.
   - (b) Remove sutures around chest tube, holding the chest tube in place.
   - (c) Drain should be removed during Valsalva maneuver or expiration.
   - (d) Pull the tube out quickly while covering the insertion site with Vaseline gauze.
   - (e) Place folded 4 × 4 gauze over the petrolatum gauze and cover with adhesive tape.
   - (f) Order a follow-up chest X-ray.

# Scoliosis Exposure

**J. Alexander Holbert, W. Fred Hess, and Mary Jane Reed**

**Key Points**

1) Anterior spinal exposures depend on the level which needs to be released. Exposures may require thoracotomy, laparotomy or thoracoabdominal with division and reconstruction of the diaphragm.
2) Thoracic: Confirm rib level with palpation of the second rib externally beneath the scapula.
3) Thoracoabdominal: Confirm total rib number preoperatively and rib level by counting from the bottom up. The diaphragm must be divided radially to preserve the phrenic nerve branches.
4) If necessary, disarticulate the rib from the costal cartilage using a periosteal elevator.
5) Lumbar: Stay anterior to the psoas to avoid the hyper-vascular retropsoas space. Identify ureter and protect from the operative field.

Coordination between the spine surgeon and exposure surgeon is imperative in preparing the patient and family for the procedure. Preoperative preparation with a review of history and physical, testing, and radiologic imaging is essential. Reviewing imaging focusing on adjacent vasculature and structures to the operative

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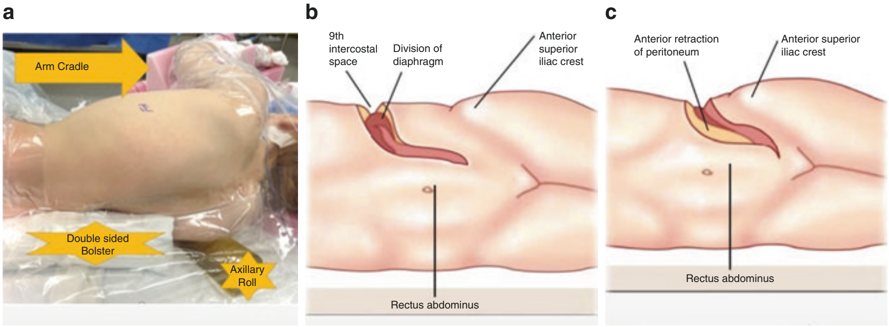
spinal objective can help prepare the exposure surgeon. Evoked potential monitoring may be deemed necessary and should be checked after positioning. It is helpful for both surgeons to be present during preoperative positioning. Cell saver is often employed.

1. Exposure of thoracic spine only:
    a. This a postero-lateral thoracotomy. Additional positioning augmentation of the spine is not necessary due to the convexity of the scoliosis. (Fig. 1a).
    b. The patient is positioned in lateral decubitus with padding and taping on a radiolucent operative table with the convex side of the spine up. (Fig. 1a).
    c. The incision is made at the rib of the most proximal discectomy planned. For example, if T6–T11 is the operative focus, then the seventh rib would be resected. If instrumentation is planned, a more proximal rib is beneficial.
    d. After dividing the latissimus dorsi muscle, release the potential space posterior to the serratus for a muscle-sparing approach.
    e. After mobilizing the potential space, a hand can be placed under the scapula to confirm rib level. The second rib is usually the most proximal that can be externally palpated.
    f. The rib is dissected subperiosteally. (Fig. 2a).
    g. The rib is then cut anteriorly for the extent of the incision and either disarticulated or cut posteriorly.
    h. The pleura can be released through the base of the rib bed, and the chest is entered. Verify rib resected by counting intrathoracically from which the first rib can be felt. Rib spreading retractor is then placed—Finochietto or other self-retaining systems (Omni or Burkholder).
    i. The lung is then deflated or packed superiorly. The pulmonary ligament may need to be divided to allow the lung to be retracted from the operative field. Care should be taken as the inferior pulmonary vein lies in close proximity.

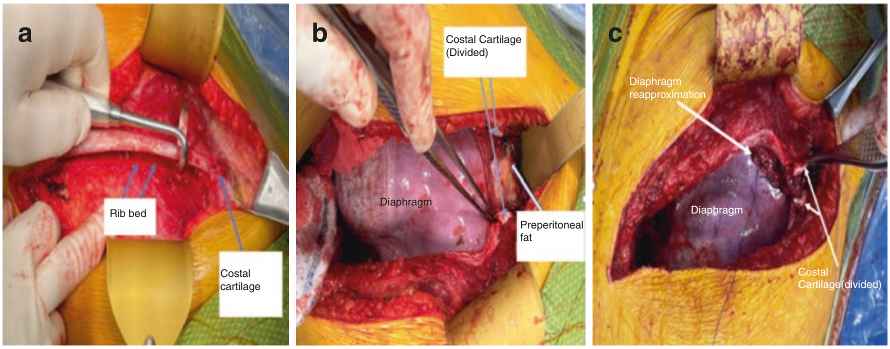

a
Arm Cradle
Double sided Bolster
Axillary Roll
b
9th intercostal space
Division of diaphragm
Anterior superior iliac crest
Rectus abdominus
c
Anterior retraction of peritoneum
Anterior superior iliac crest
Rectus abdominus

Approach to Thoracic Spine

Approach to Thoracoabdominal Spine

Approach to Lumber Spine

**Fig. 1** Surgical approaches to (**a**) the thoracic spine, (**b**) the thoracoabdominal spine, and (**c**) the lumbar spine. (Source: Christopher Coppola, MD and Fred Hess, MD)


a
Rib bed
Costal cartilage
b
Costal Cartilage (Divided)
Diaphragm
Preperitoneal fat
c
Diaphragm reapproximation
Diaphragm
Costal Cartilage(divided)

Subperiosteral rib dissection　Identifying preperitoneal plane　Closure of diaphragm

**Fig. 2** Thoracoabdominal spine exposure. (**a**) Dissection of rib. (**b**) Division of costal cartilage. (**c**) Closure of diaphragm. (Source: W. Fred Hess, MD)

   j. An internal thoracoplasty (cutting the rib at the rib angle and disarticulating the head or cutting at the level of the transverse process) can then be done if more space is required or if deemed clinically needed (this also provides an excellent source of autogenous bone graft). Take care not to injure the intercostal bundle during dissection of rib.
   k. The pleura is then released in the mid-lateral line of the vertebra, and segmental vessels are doubly ligated. The pleura can then be dissected anteriorly around the vertebral bodies. A lipped malleable retractor can then be placed in this interval protecting the great vessels while providing retraction.
   l. After the spinal procedure is complete, hemostatic agents such as thrombin-soaked gel foam or Floseal ™ may be utilized to seal the disc spaces. Bone wax maybe helpful in decreasing bleeding from bone perforators.
   m. Reapproximate pleura over the spine.
   n. Place chest tube as a drain under direct visualization and position postero-lateral.
   o. Have Anesthesiologist apply positive pressure to ensure lung inflates fully.
   p. Administer intercostal block at the resection level, including two above and two below the rib resection. (Also, at any thoracoplasty levels).
   q. Place paracostal sutures if necessary.
   r. Reapproximate the rib bed.
   s. Ask Anesthesiologist to apply and hold positive pressure to ensure there is no air leak.
   t. Close layers sequentially.

2. Exposure of thoracoabdominal exposure:

   a. Before surgery, verify the total number of ribs by reviewing imaging.
   b. Positioning is the same as previously detailed with operative field widened to include pelvis. (Fig. 1b).

c. An oblique incision is made over the planned rib to be resected ninth, tenth, or eleveth tenth (depending on the level required). Make incision one level above the disc to be resected.
d. Latissimus and external oblique muscles are released in line with the rib from the erector spinae muscle posteriorly to the lateral border of the rectus anteriorly.
e. The rib is dissected subperiosteally, the internal oblique and transversalis are released in line with the costal cartilage.
f. The rib is disarticulated from the costal cartilage anteriorly and either disarticulated or cut posteriorly. Verify correct rib resected by counting backwards from twelveth rib.
g. Enter the chest through the midline of the rib bed. Self-retaining retractor is placed. Pack lung superiorly and release the pleura in the mid-lateral line. Release the pleura down to the crus of the diaphragm. The line from the costal cartilage to the midpoint of the vertebra is then scored on the diaphragm to delineate the division.
h. The costal cartilage is then split with a scalpel in line with the rib, the preperitoneal fat is then visualized. (Fig. 2b).
i. The space between the peritoneum and the diaphragm is released. The potential space is enlarged. The diaphragm is divided radially. Tagging sutures are helpful to facilitate diaphragmatic repair and can be used for retraction of diaphragm.
j. Doubly ligate segmental vessels. (Beware of T12/L1, which is hidden under the crus of the diaphragm).
k. Dissection is continued caudally to the psoas to the distal level needed. Care is taken to remain anterior to the psoas. Knowledge of at risk structures both in the operative field and behind the retractors is imperative. Occult injury to intraperitoneal structures during aggressive retraction have been reported.
l. After spinal procedure is complete, hemostatic agents such as thrombin soaked gel foam or Floseal™ may be utilized to seal the disc spaces.
m. Reapproximate the pleura along the spine (may not be possible with instrumentation).
n. The diaphragm is reconstructed with permanent suture, taking care not to leave gaps which would become sites of diaphragmatic hernia. This can be reinforced with 2–0 Ticron with pledgets. Transversalis muscle is closed in line with the diaphragm. The costal cartilage is very helpful as a guide when reapproximating these layers. (Fig. 2c).
o. Place chest tube to re-inflate lung and position postero-laterally.
p. Ask anesthesia to apply positive pressure to ensure lung inflates fully.
q. Intercostal block at the resection level plus two levels above and below the rib resection. (Also, at any thoracoplasty levels).
r. Rib bed closure in line with the internal oblique muscle and reapproximating the costal cartilage.
s. Ask Anesthesiologist to apply positive pressure to ensure there is no air leak.
t. Close latissimus and external oblique muscles in continuity.

3. Exposure of lumbar spine only: Rarely used for scoliosis.
   a. Positioning is the same as previously detailed.
   b. The incision follows the curve of the superior iliac crest from anterior to posterior, choosing the distance between the bone and incision. The incision is centered over the level of vertebra desired. (Fig. 1c).
   c. Divide the external oblique muscle and aponeurosis in the direction of fibers from the edge of the rectus posteriolaterally to the front edge of the latissimus dorsi.
   d. Divide across the internal oblique muscle fibers.
   e. Carefully divide the transversalis muscle, preserving the peritoneum, pushing it anteriorly, and bluntly dissecting it off of the underside of the flank body wall. The dissection is easier in the posterior portion of the transversalis muscle, where it is not so densely adherent to the peritoneum. Repair any holes created in the peritoneum with an absorbable suture taking care not to injury underlying structures.
   f. Continue mobilizing the peritoneum and viscera within, working toward the posterior edge until the spine comes into view. Often there is a layer of retroperitoneal fat in this plane that facilitates the dissection.
   g. Dissection needs to be carried anteriorly to the psoas down onto the spine.
   h. If the incision area is L5-S1, the spermatic cord or round ligament may be encountered and mistaken for the ureter. The ureter is usually located along the psoas and deeper transversing the iliac vessels.
   i. Avoid dissecting the hypervascular retropsoas space.
   j. Retroperitoneal structures, namely ureter, aorta, and vena cava branches are carefully identified and retracted anterior, suture ligating the segmental vertebral venous and arterial branches as necessary. The iliac vessels may need to be circumferentially dissected and encircled with vessels loops so they can be manipulated from the vertebrae.
   k. If dissection is carried down to the L4/5-L5/S1 disc level, the iliolumbar vein must be identified and protected or ligated.
   l. After the spinal procedure is complete, hemostatic agents such as thrombin-soaked gel foam or Floseal™ may be utilized to seal the disc spaces.
   m. Close layers of the abdominal wall, taking care not to injure the ureter or viscera.
   n. Ipsilateral pedal pulses should be assessed before the breakdown of the sterile field.

## Study Questions

1.) Which one of the following structures should be identified and retracted out of operative field during lumbar spinal approach?
   a. Genitofemoral nerve.

b. Iliolumbar vein.
c. Ureter.
d. Round ligament.

Answer: (c.) Ureter. The ureter is usually encountered medial to psoas crossing over iliac vessels. It most often adheres to the reflected peritoneal structures once in the retroperitoneal space. The genitofemoral nerve should be identified but will remain adherent to the underlying psoas muscle. The iliolumbar vein is usually ligated and divided during dissection of L4 to help mobilize the distal aorta and vena cava. The round ligament is a suspendatory ligament of the uterus, it is dorsal to the retroperitoneum and caudal to the usual operative field of lumbar dissection. It can be confused with the ureter in slender patients however it does not peristalsis.

2.) Which of the following considerations is FALSE during thoracoabdominal approach to spine.

a. When dividing the diaphragm, the incision should be through the center of the muscle to allow for adequate tissue for closure.
b. The pleural should be closed over the spinal repair if possible.
c. The costal cartilage is useful as guide to both preperitoneal plane at the beginning of dissection as well as during closure of diaphragm and muscular layers.
d. The T12/L1 segmental vessels maybe obscured by the crus of the diaphragm.

Answer: (a.) (FALSE) When dividing the diaphragm, the incision should be through the center of the muscle to allow for adequate tissue for closure. The innervation of the diaphragm by the phrenic nerve is from central to periphery, therefore incision of diaphragm during thoracoabdominal approach should be along the periphery. The pleural provides some constrictive hemostatsis and should closed over repair if possible. The costal cartilage at medial edge of rib is helpful as a landmark as the beginning of the preperitoneal space that can be developed laterally. The crus of the diaphragm at T12/L1 can obscure the segmental vessels making the vessels more difficult to identify.

## Further Reading

Bartley CE, Yaszay B, Bastrom TP, Shah SA, Lonner BS, Asghar J, Miyanji F, Samdani A, Newton PO. Perioperative and delayed major complications following surgical treatment of adolescent idiopathic scoliosis. J Bone Jt Surg Am. 2017;99:1206–12.

Watkins RG III, Watkins RG IV. Surgical Approaches to the Spine. 3rd ed. Springer; 2015.

# Hydrocephalus and CSF Diversion

**Angel G. Chinea, George Jallo, and Nir Shimony**

**Key Points**

1) Hydrocephalus (HCP) is a pathologic condition that results from excess accumulation of cerebrospinal fluid (CSF) within the ventricular system leading to increased intracranial pressure.
2) Early recognition and treatment of this entity is important to prevent death or permanent disability.
3) CSF diversion procedures are varied, the most common being ventriculoperitoneal shunt (VPS) and endoscopic third ventriculostomy (ETV).

1. Background
    a. Epidemiology: incidence of 0.9–1.8/1000 live births with estimated prevalence of 1–1.5% [1, 2].

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b. Etiologies:
    i. Congenital
        1. Myelomeningocele—independent of the presence of a Chiari II malformation.
        2. *Aqueductal stenosis—primary atresia or narrowing of the aqueduct of Sylvius *(can also be acquired secondary to scarring from intrauterine infection or germinal matrix hemorrhage).
        3. Dandy–Walker malformation—atresia of foramina Luschka and Magendie.
        4. Chiari I malformation—fourth ventricular outflow obstruction (It is paramount to rule out a secondary Chiari due to untreated hydrocephalus, before declaring the hydrocephalus secondary to fourth ventricular outflow obstruction).
        5. Hydranencephaly.
        6. Holoprosencephaly.
        7. Porencephaly.
        8. Schizencephaly.
        9. VACTERL-H.
        10. X-linked inherited.
        11. Unexplained cause, other.
    ii. Acquired
        1. Infectious
            a. Cryptococcus.
            b. Tuberculosis.
            c. post-meningitis—usually bacterial.
        2. Hemorrhagic
            a. Intraventricular germinal matrix hemorrhage (more common after IVH grade 3 or 4).
            b. Post-subarachnoid hemorrhage (SAH).
        3. Mass occupying lesions
            a. Tumors—usually intraventricular or tectal region lesions, however parenchymal lesions can also cause HCP based on their location (diencephalon, pons, suprasellar, etc.)
            b. Non-neoplastic—AVMs, aneurysms, vein of Galen malformation, intracranial cysts, and others depending on location.
            c. Postoperative—can be seen in up to 20% of pediatric patients following posterior fossa surgery [1, 2].
        4. Spinal cord tumor—mechanism is not well understood.

2. Pathophysiology:
    - a. Ventricular anatomy: The ventricular system is made up of two lateral ventricles, a third ventricle, and a fourth ventricle connected via small pathways and sharing a common outflow tract. They are fluid filled spaces lined by ependymal cells and contain the choroid plexus where CSF is made. The two C-shaped lateral ventricles span the length of each cerebral hemisphere and are separated by the midline septum pellucidum. Each lateral ventricle has an interventricular foramen of Monro within their frontal horns that connect them to the third ventricle ventrally. The slit-like third ventricle lies between both thalami and is connected to the fourth ventricle posteriorly via the cerebral aqueduct of Sylvius. The fourth ventricle is surrounded by the brainstem anteriorly and cerebellum posteriorly and contains the medial foramen of Magendie and two lateral foramina of Luschka that serve as the ventricular outflow tracts into the cisterna magna at the base of the brain. CSF within the subarachnoid space can then flow around the spinal cord inferiorly toward the lumbar cistern and intracranially within the cerebral cisterns and surface of the brain.
    - b. CSF Physiology: Cerebrospinal fluid is a clear, sterile fluid that surrounds the brain and spinal cord and thought to play a role in both nutrient exchange and mechanical “cushion” of CNS structures. Most of it (80%) is produced by the choroid plexuses and circulates the CNS until being absorbed by the arachnoid granulations lining the dural sinuses and into the venous circulation [1, 3].
3. Clinical features:
    - a. Types:
        - i. Obstructive: blockage of normal CSF pathways anywhere from the foramen of Monroe to the fourth ventricular outlet tracts resulting in fluid accumulation proximal to obstruction. Atresia of foramen of Monroe will cause enlargement of one lateral ventricle, whereas thalamic or third ventricular lesion is expected to cause enlargement of both lateral ventricles. Obstruction of the Aqueduct of Sylvius will result in “triventricular” enlargement on imaging and all 4 ventricles will be enlarged with outflow obstruction.
        - ii. Communicating: impaired CSF reabsorption at the arachnoid granulations. This is thought to occur either from scarring or decreased reabsorption capacity in the setting of degrading blood products or increased protein/cells in the CSF, respectively.
    - b. Presentation:
        - i. Acute HCP—relatively rapid neurologic decline that occurs when compensatory physiologic mechanisms are unable to offset the effects of CSF accumulation. Timing can vary from just hours in the older patient with

rigid cranial vault to weeks in the neonate with open fontanelles and unfused cranial sutures. This presentation is usually fatal or will lead to irreversible neurologic injury if unrecognized or treatment is delayed.*

    ii. Chronic HCP—usually seen with incomplete obstruction or relatively slow accumulation of CSF that allows for compensatory mechanisms to prevent death. Patient may present with macrocephaly generally >98th percentile, spontaneous CSF leak, vision problems or gaze palsies, and findings on imaging such as "empty sella," callosal atrophy, or "beaten copper" cranium on X-rays. Younger children can also present with failure to thrive and neurocognitive delay, increasing head circumference crossing percentiles, delayed fontanelle closure, and sutural diastasis [1, 2].
    iii. "Arrested" HCP—This term has been used synonymously with compensated hydrocephalus and refers to a patient with imaging findings of ventriculomegaly but HCP symptoms at presentation are either mild or resolved. Criteria to satisfy this diagnosis include slightly enlarged ventricle caliber, normal head growth curve, and normal neurocognitive and psychomotor development in the absence of a shunt [1, 2].
    iv. Ventriculomegaly—enlarged ventricle size on imaging can be suggestive but not in itself diagnostic of hydrocephalus. Cerebral parenchymal volume loss after anoxic injury or developmental anomaly, for example, will give the appearance of ventricle enlargement but these patients do not require treatment as ICP levels are normal.

* from a practical perspective the information pertaining to diagnosis and treatment below will be mostly referring to the management of acute HCP.

4. Diagnosis

    a. Signs and symptoms—dependent on whether cranial sutures are open or fused.

        i. Infants (<2 y/o)—tense or bulging fontanelle, splayed sutures, macrocephaly, increasing head circumference crossing percentiles, scalp vein engorgement, apneic spells, and bradycardic episodes. They can also exhibit features of raised ICP (abducens palsy, irritability, etc.) as discussed below.
        ii. Older patients—generally limited to those of increased ICP such as nausea, vomiting, headaches, lethargy, irritability, and gait difficulty. Specific findings can include upward or CN 6 palsies, pupillary dilatation, or papilledema.

    b. Radiographic Studies:

        i. Cranial U/S—as long as the anterior fontanelle is open it is useful in neonates for diagnosis of ventriculomegaly, germinal matrix hemorrhage, and intracranial cysts and for tracking progression of ventricle size over time.

    ii. MRI brain—the most sensitive and specific modality to delineate brain anatomy and identify pathology causing HCP.
    iii. CT—is useful for identifying enlarged ventricle size and for gross comparison if prior films available. Generally limited to the emergency setting due to long-term risks of radiation exposure in the pediatric population.

5. Treatment
    a. Goals: CSF diversion can be accomplished by either shunting excess CSF into an extra-ventricular cavity or bypassing site of blockage by taking advantage of the local anatomy (ETV). In cases of HCP secondary to mass effect, the primary focus should be on resecting or debulking the offending lesion and challenging the need for permanent shunting by using temporary measures. Similarly, temporary CSF diversion is also used in neonates with germinal matrix hemorrhage as up to 80% of these will not need a permanent shunt.
    b. Available options:
        i. Permanent
            1. Ventriculoperitoneal shunt—most commonly used for treatment of HCP around the world. It works by directly shunting fluid away from the ventricle into the peritoneal cavity. Its components include a ventricular (proximal) catheter, reservoir, valve, and peritoneal (distal) catheter connected in series. Reservoirs can be attached to the proximal catheter or be a component of the valve itself. There are multiple valve types and brands but are generally divided into programmable or non-programmable (fixed-pressure) valves.
            2. Other shunt types—similar to VPS but with different terminus.
                a. Ventriculopleural—distal catheter is placed in the pleural space, usually in the anterior axillary line between fourth and fifth ribs. It is controversial to use it in young kids (less ability to absorb large fluid quantity from the pleural cavity).
                b. Ventriculoatrial—distal catheter is inserted into the internal jugular vein and terminates in the right atrium. This is the most popular option in children after the ventriculoperitoneal shunt.
                c. Rarely utilized but alternative options include ventriculo-ureteral, ventriculo-gallbladder, and ventriculo-sagittal sinus shunt.
            3. Endoscopic third ventriculostomy—This procedure works for treatment of obstructive hydrocephalus by creating an ostomy in the premammillary membrane at the floor of the third ventricle, therefore diverting CSF into the interpeduncular cistern. The main benefit of ETV is that it does not require a foreign object to be left inside the body.
            4. Choroid plexus coagulation—infrequently used procedure for treatment of HCP, however is currently being performed in certain specialized centers in conjunction to ETV for infants less than 2 years (usually less than 1 year of age). The benefit of it is still being studied [4, 5].

   ii. Temporary

      1. External ventricular drain (EVD)—A catheter similar to a proximal shunt catheter is inserted into the lateral ventricle using same techniques as for VPS surgery. This can be performed in the OR or at the bedside in the emergent setting. The catheter is tunneled away from the incision and connected to an external collecting system with the capacity for ICP monitoring, making it useful to challenge need for shunting as well as for management of severe TBI.
      2. Subgaleal shunt—Similar to VPS however the proximal catheter is connected to a reservoir and distal catheter terminates beneath the galea of the scalp. Valves are generally not used and function can be tracked by observing presence of subcutaneous fluid collection. These are used mostly for neonates following germinal matrix hemorrhage who may not require permanent shunting.
      3. Ventricular puncture—this can be used as an emergent or temporizing procedure in small or preterm neonates who are unable to undergo shunt surgery. This is performed at the bedside in the NICU and serial punctures may be required in select cases over a period of time.

c. Description of procedure (in brief):

   i. VPS—The patient is positioned with the head turned away from the side of planned ventricular access. The catheter entry site is identified based on external landmarks or through platform-based navigation systems. The most commonly used landmarks are Kocher's point (frontal) and Frazier's point (occipital), however there are a few others as illustrated in the figures below (Figs.1, 2, 3, and 4). The patient is then prepped from the head down to the waistline and draped. A semilunar incision is made at the scalp and burr hole created. At this point it is this author's preference to

| | Obstructive | Communicating |
|---|---|---|
| Congenital | 1. Chiari I<br>2. Aqueductal stenosis<br>3. Dandy-Walker | 1. Myelomeningocele<br>2. Hydranencephaly<br>3. Holoprosencephaly<br>4. Porencephaly<br>5. Schizencephaly<br>6. VACTERL-H<br>7. X-linked inherited<br>8. Unexplained, other |
| Acquired | 1. Tumors<br>2. Intracranial cysts<br>3. AVMs & other vascular lesions | 1. Cryptococcus<br>2. Tuberculosis<br>3. Post-meningitis<br>4. Germinal Matrix hemorrhage<br>5. SAH<br>6. Post-operative |

**Fig. 1** Chart of HCP etiologies divided by type

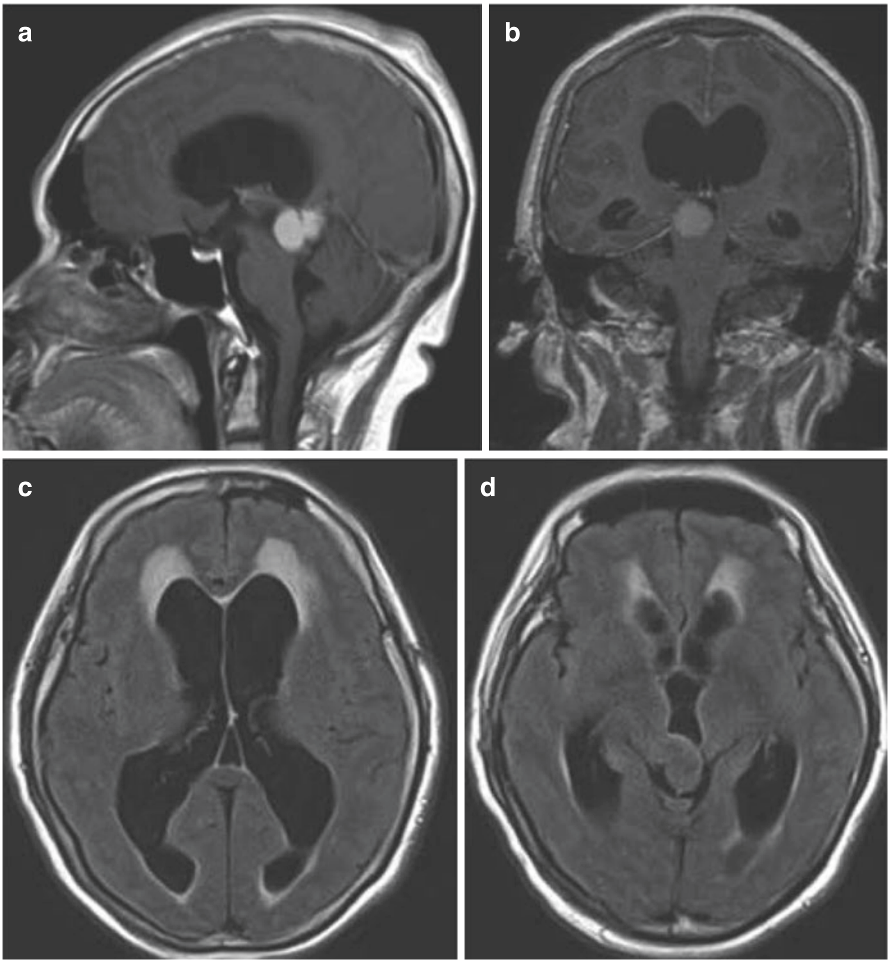

**Fig. 2** Example of obstructive hydrocephalus. (**a**–**d**) include sagittal and axial MR images showing "triventricular" hydrocephalus secondary to pineal region mass causing obstruction of the cerebral aqueduct of Sylvius [6]. Source: Image courtesy of Ling & Tiruchelvarayan—A Review of Endoscopic Treatment of Hydrocephalus in Paediatric and Adult Patients. *Proceedings of Singapore Healthcare*

tunnel the distal catheter prior to creating the durotomy and this can be done either from the head caudally or from the abdomen cranially. (Sometimes a small retroauricular incision is needed for complete tunneling of frontal location shunts.) The ventricle is then accessed and after confirming spontaneous CSF flow is connected either to a reservoir-valve series or integrated valve which is then connected to the distal catheter. Distal flow is confirmed and the distal catheter is then inserted into the peritoneum either through a small open laparotomy or laparoscopically through small stab incision with possible general surgery assistance.

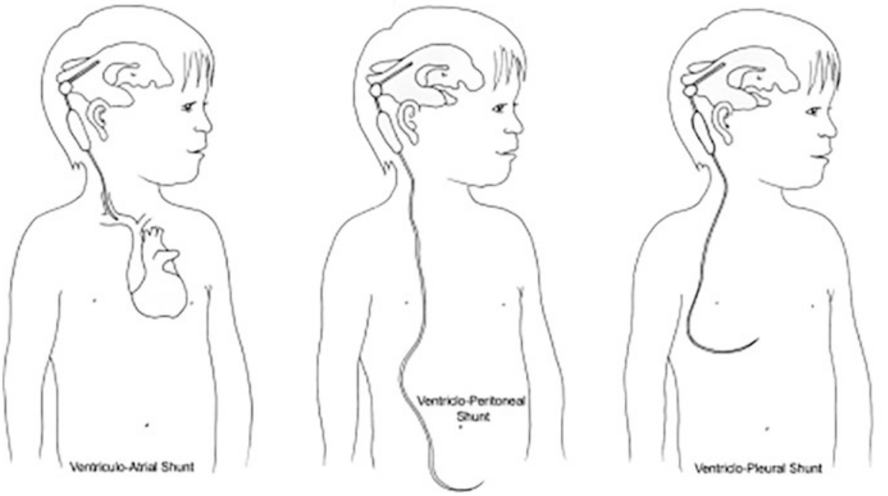

**Fig. 3** Schematic drawing of different CSF shunt types. These images represent a simple schematic summary of the three most commonly implanted ventricular shunt types [7]. From the left: ventriculo-atrial shunt, ventriculo-peritoneal shunt, and ventriculo-pleural shunt. Source: Image courtesy of Sally Gooden illustrator for LEEDs teaching hospitals. *Hydrocephalus and Shunts, information for patients*

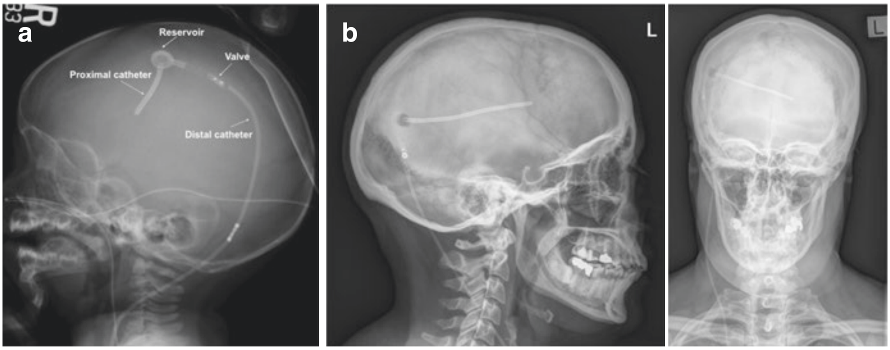

**Fig. 4** Plain radiographs of shunts with different ventricular catheter trajectories. (**a**) Lateral, plain X-ray depicting a frontal approach (Kocher's point) ventricular catheter with its labeled components [8]. (**b**) lateral and AP X-ray films showing an occipital approach (Frazier) ventricular catheter [9]. Source: (**a**) image courtesy of Maller et al., *Seminars in ultrasound, CT, and MR*. (**b**) Image courtesy of Andrew Murphy, *Radiopaedia*

ii. ETV—The patient is positioned supine with head elevated in a neutral position. Ventricular access is usually planned using Kocher's point as an entry site, with some modifications that sometimes need to be taken based on the patient's specific ventricular anatomy. Navigation if available can be used to optimize the trajectory to target and maximize efficiency and safety. After prepping and draping the patient, a small incision is made in the scalp and a burr hole created over the planned entry site. A small durotomy is made and ventricular access is gained with a sheath covered trocar. Once the sheath secured to the scalp a ventriculoscope is advanced and passed through the foramen of Monroe into the third ventricle. Once the thin premamillary membrane is identified it is important to be able to determine location of basilar artery and mamillary bodies to avoid injury during ostomy creation. There are several techniques that can be employed to create a hole in the floor of the third ventricle. The defect can be expanded using balloon dilatation or through hydrodissection alone.

d. Postoperative care:

    i. Patients are observed for a period of 24–48 hrs following shunt or ETV procedures and this can be done in a med-surg floor if procedure is uncomplicated. A postoperative rapid sequence MRI can be obtained to confirm catheter location and as a new baseline for future reference.

e. Complications:

    i. There are common complications related to shunt surgery that are important to recognize as these may lead to shunt malfunction or rapid neurologic decline.

        1. Obstruction—most common cause of malfunction with incidence of 35–40% within the first year of placement. Proximal catheter is the most common site of blockage [1, 2, 10].
        2. Infection—incidence is reported to be 8–15% and there is an elevated risk during the first 6–12 months following surgery [10]. The most common organism(s) are *S. epidermidis* and *S. aureus,* followed by Gram negative species [1, 2, 10]. While there is data to suggest that antibiotic impregnated catheters reduce the incidence of shunt infection, the only intervention that has consistently shown an improvement is implementation of institution-specific perioperative protocol for shunt placement [10].
        3. Disconnection or “break”—usually seen with old shunt systems or in growing children.
        4. Skin erosion—mostly in neonates or sick children with poor nutritional status. Can be indicative of silicone allergy.
        5. Peritoneal pseudocyst—limited to ventriculoperitoneal shunts. Occurs secondary to chronic inflammation thought by many to be indicative of indolent infection or colonization of catheter tip by bacteria [1, 10].

## Additional Notes

Hydrocephalus is a neurologic condition that has been recognized for millennia, however it was largely considered terminal until the late nineteenth and early twentieth century when aseptic technique and better understanding of human pathophysiology began to yield favorable surgical outcomes. Further advancements in the field of medicine and technology over the subsequent one hundred years have resulted in the development of surgical interventions that are extremely safe and effective, thereby eliminating mortality as a common procedural complication. The responsibility of the pediatric physician to prevent serious death or disability lies in early recognition of this pathology in order to provide timely treatment.

There are multiple studies and diagnostic criteria that utilize radiographic findings to try to predict likelihood of HCP (such as Evan's ratio, empty sella, third ventricle ballooning, etc.) but ultimately the diagnosis can only be made by considering these findings in conjunction with clinical presentation and physical examination. If clinical suspicion is marginal, temporary measures such as an EVD trial can be employed prior to subjecting the patient to permanent shunting. All patients that are diagnosed with HCP and deemed surgical candidates should be evaluated for an ETV, which can include specific MRI sequences to assess for CSF flow dynamics. Another tool used by pediatric neurosurgeons is the ETV success score (ETVSS), which takes into account the patients age, HCP etiology, and shunt history to predict ETV success (See image below).

Treating HCP in the developing world remains a clinical challenge due to limited access of certain populations to routine postnatal care or qualified surgical facilities. ETV has become an attractive option as no implants are required and postoperative complications are low, however this requires specialized equipment and more careful patient selection to ensure success. VP shunting remains the most common procedure for treating HCP in the developing World and fixed-pressure valves tend to be favored due to lower risk of malfunction and less maintenance.

## Study Questions

1) A 7 y/o male with new diagnosis of Chiari I malformation was referred to the hospital due to enlarged ventricles on brain imaging. Which of the following findings would be concerning for developing hydrocephalus?

   a. Headaches
   b. Abnormal gait
   c. Vision decline with papilledema
   d. All of the above

   Answer: d—All of the above. Hydrocephalus is a clinical diagnosis that occurs secondary to abnormal CSF accumulation resulting in increased ICP and can be due to ventricular outflow obstruction such as in this case. Ventriculomegaly is a radiographic finding that, while it is usually associated with hydrocephalus, is not diagnostic in the absence of clinical symptomatology. Benign macrocrania is an example of such where patient have above average head circumference and ventricle caliber but are asymptomatic and shunt placement is not indicated. Likewise, it is important to recognize symptoms of elevated ICP and associated conditions as some patients may present with normal sized ventricles on imaging.

2) A patient with obstructive hydrocephalus secondary to a colloid cyst blocking the interventricular foramen of Monroe should be treated with:

   a. Ventriculoperitoneal shunt
   b. Endoscopic Third Ventriculostomy (ETV)
   c. Cyst resection with EVD trial
   d. a & c

   Answer: d—a & c. A patient with an obstructive lesion at the foramen of Monroe will cause CSF accumulation within one or both of the lateral ventricles depending on size. An ETV would not treat this patient's HCP as the ostomy is created distal to the obstruction. Most surgeons would advocate for (c) as resecting the lesion will usually open up the CSF pathways and placement of temporary CSF shunt intraoperatively is a reasonable and safe method of confirming total resolution of obstruction in the acute postoperative period. (a) Permanent VPS placement up front is the less preferred option but is a reasonable alternative to treat hydrocephalus in a patient that otherwise would not tolerate or opts against the riskier cyst resection.

3) Which of the following conditions cause transient HCP which may not require permanent CSF shunt?

   a. Aqueductal stenosis
   b. X-linked hydrocephalus
   c. Germinal matrix hemorrhage (GMH)
   d. Vein of Galen Malformation

   Answer: c—Germinal Matrix Hemorrhage. Hydrocephalus with this pathology is typically associated with grade 3&4 GMH and thought to be related to decreased CSF reabsorption due to degrading blood products. In most centers, treatment of hydrocephalus usually includes subgaleal shunts or serial ventricular taps as less than half of patients will develop long-lasting hydrocephalus requiring permanent shunt.

## References

1. Greenberg. Handbook of Neurosurgery, ninth edition. Hydrocephalus and Cerebrospinal Fluid. Thieme. 2020:396–453.
2. Jallo, et al. Handbook of Pediatric Neurosurgery. Surgical Management of Hydrocephalus & Treatment of Hydrocephalus in the Developing World. Thieme. 2018:297–311.
3. Mortazavi, et al. The ventricular system of the brain: a comprehensive review of its history, anatomy, histology, embryology, and surgical considerations. Childs Nerv Syst. 2013;
4. Kulkarni, et al. Endoscopic third ventriculostomy and choroid plexus cauterization in infants with hydrocephalus: a retrospective Hydrocephalus Clinical Research Network study. JNS pediatrics. 2014;14:224–9.
5. Souweidane MS. Editorial: Combined choroid plexus coagulation and endoscopic third ventriculostomy: is North America ready? JNS. 2014; https://doi.org/10.3171/2014.4.PEDS1450.

6. Ling J, Tiruchelvarayan R. A Review of Endoscopic Treatment of Hydrocephalus in Paediatric and Adult Patients. Proceedings of Singapore Healthcare. 2013;22:203–12.
7. LEEDs teaching hospitals. Hydrocephalus and Shunts; information for patients, *Illustrator-Sally Gooden*. NHS Trust. 2nd edition version 1. 2016, reviewed 2018.
8. Maller V, et al. Imaging of Ventricular Shunts. Semin Ultrasound CT MR. 2016;37:159–73.
9. Murphy et al. Shunt Series. *Radiopaedia*. Last Revision 2020.
10. Paff, et al. Ventriculoperitoneal shunt complications: A review. Interdisciplinary Neurosurgery. 2018;13:66–70.

# Circumcision

**Christopher P. Coppola**

**Key Points**

1) Necessity for routine male circumcision is controversial, and there is a wide variation of circumcision rate between different cultures and geographic regions.
2) Potential benefits of male circumcision are reduced urinary tract infections (UTI), reduced transmission of sexually transmitted diseases, and reduction in incidence of penile cancer [1].
3) Postoperative bleeding is the most common complication, but providers also need to be vigilant for more serious complications such as injury to the glans or urethra and retained in situ ring device causing urinary obstruction or ischemia to the glans.

Routine male circumcision is a widely debated topic. It is frequently performed soon after birth in the United States in approximately 60% of male newborns [2]. It is currently not routinely indicated in all male newborns and should be considered elective. The 2007 statement from the American Academy of Pediatrics states that the potential health benefits outweigh the risks, so the procedure should be made available to parents of all male newborns who choose it, after they have received proper counseling [1]. Many parents opt for routine neonatal circumcision for religious, cultural, or hygienic reasons. Potential conditions of the foreskin include phimosis, paraphimosis, and balanitis. Potential health benefits of male circumcision are prevention of urinary tract infections, some sexually transmitted diseases,

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and penile cancer [1]. It is safe and acceptable for parents to choose not to have their child circumcised, unless there is a condition causing obstruction or recurrent infections.

1. Pathophysiology: Most males are born with fusion of the underside of the prepuce to the glans; the foreskin often hides the meatus. This fusion prevents the foreskin from being retractable. The natural process of separation of the prepuce and the glans occurs over months but may take several years. By puberty, the foreskin should be completely retractable.
2. Contraindications:
    a. Penile abnormalities: hypospadias/epispadias, chordee, ambiguous genitalia.
3. Procedure:
    a. Circumcision devices: These are primarily used in newborns. Common devices include: the Gomco clamp (Goldstein Medical Company, NY), Plastibell (Hollister Incorporated, Libertyville, IL), or Mogen clamp ("shield" in Hebrew) (Fig. 1).
        i. General procedure guidelines:
            1. Adequate anesthesia should be obtained: Tylenol, local anesthetic, etc.
            2. Antisepsis: sterile technique.
            3. Adhesions between the glans and the underside of the foreskin are bluntly separated.
        ii. Gomco clamp—most common device:
            1. A dorsal slit is made to allow for the bell device to fit over the glans and under the foreskin.
            2. The bell and overlying foreskin are brought through the clamp and the yoke is tightened. The excess prepuce is then excised using a scalpel.
            3. The device is removed after about 3–5 min and antibiotic ointment applied.
        iii. Plastibell—uses strangulation of cut edge of foreskin with a ligature around an in situ plastic ring to remove foreskin.
            1. Dorsal slit is made to allow for Plastibell device to fit over glans and within the foreskin.
            2. A string is tied around a groove in the Plastibell near the glans. The excess skin is trimmed and the handle of the Plastibell snapped off the bell. The remaining Plastibell and foreskin will slough off in the next 1–2 weeks.
            3. In situ ring devices like the Plastibell should only be used when there is access to postoperative surgical follow-up. There is a 2–5% complication rate, including bleeding, edema, and retained ring with ischemia of the glans.

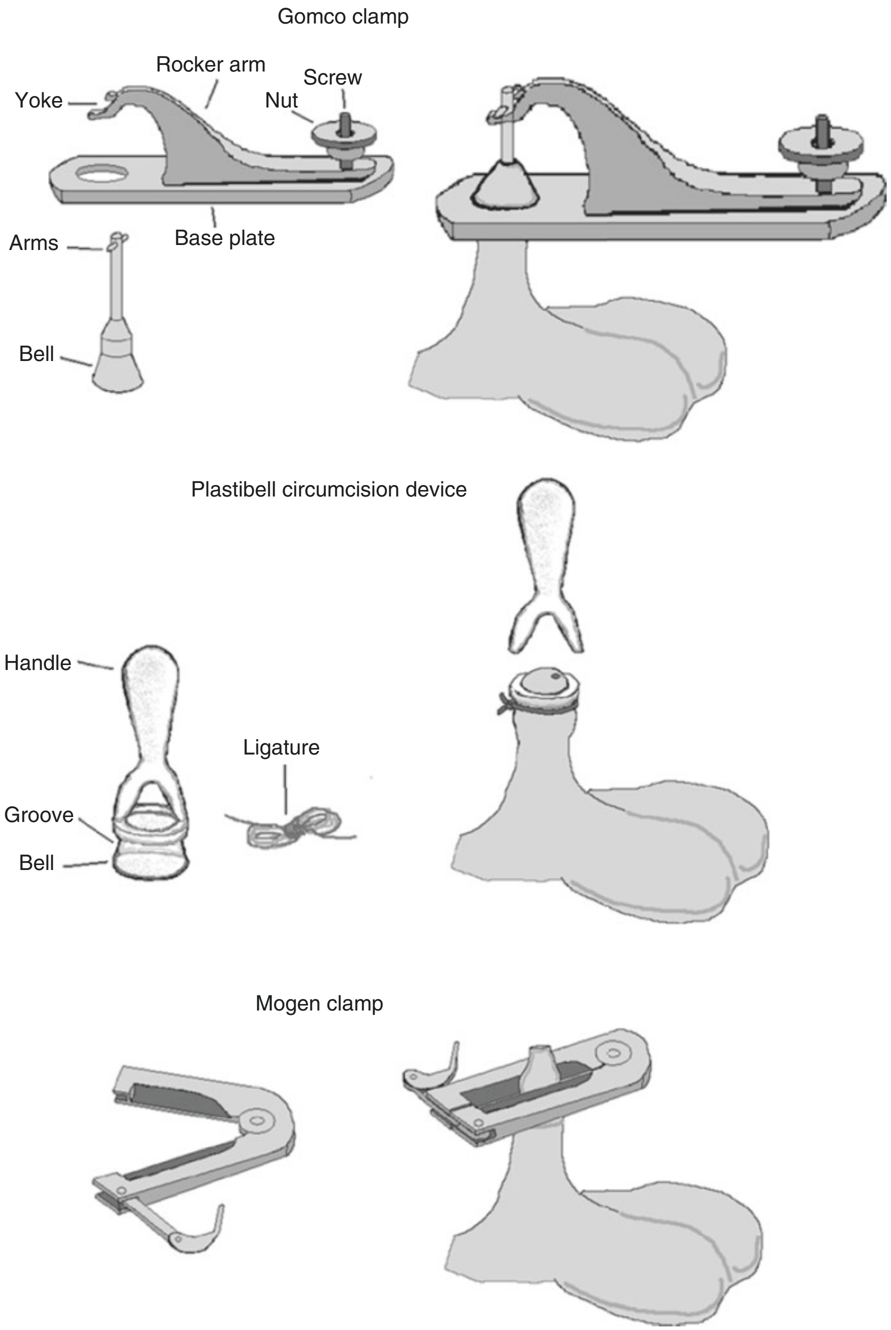
Gomco clamp
Rocker arm
Screw
Yoke
Nut
Arms
Base plate
Bell
Plastibell circumcision device
Handle
Ligature
Groove
Bell
Mogen clamp

**Fig. 1** Devices used to perform circumcision. The three most common devices used to perform circumcision are the Gomco clamp, the Plastibell circumcision device, and the Mogen clamp (Source: Christopher Coppola)

  b. Sleeve Resection: performed on older patients in the operating room.
      i. A general anesthetic is used, the patient is prepped in the typical sterile manner, and the adhesions between the glans and foreskin are bluntly taken down.
      ii. The prepuce is placed in its usual position and marked at the level of the coronal sulcus.
      iii. The prepuce is then retracted, and an incision is made circumferentially along the base of the glans. Dissection is carried down to Buck's fascia.
      iv. A second incision is made along the marked foreskin and this cuff of skin is excised.
      v. Care should be taken not to injure the urethra or ventral frenulum.
      vi. The remaining cuff of skin is then approximated with interrupted absorbable sutures to the skin just below the glans at the coronal sinus.

4. Complications:
    a. Persistent bleeding
    b. Urethral or glans injury
    c. Adhesions or scarring
    d. Too little or excessive excision of skin
    e. Edema with retained in situ ring and ischemia of glans
    f. Urinary obstruction

5. Post-op care:
    a. Some edema, bleeding, and crusting should be expected in the first few days.
    b. Antibiotic ointment or a petroleum-based ointment can be applied to the area.
    c. Penis should be gently cleansed with soap and water as needed.

Question 1: Which of the following conditions is a contraindication to performing circumcision?

a. Phimosis
b. Urinary Tract Infection
c. Balanitis
d. Hypospadias

Answer: (d.) Hypospadias. Hypospadias is a contraindication to circumcision. The foreskin may be necessary to provide material to repair the urethra during a future operation for hypospadias. If there is hypospadias, the child should be referred to a Pediatric Urologist, and circumcision should be deferred until the plan for hypospadias repair has been determined. Circumcision is a treatment for phimosis, recurrent urinary tract infections, and balanitis. These conditions may need acute treatment and evaluation such as urine culture, antibiotics, urgent dorsal slit to treat urinary obstruction, and evaluation for upper urinary tract abnormalities as a source of infection.

Question 2: Which method of circumcision has the risk of retained in situ ring causing postoperative edema and/or urinary obstruction?

a. Mogen clamp
b. Gomco camp
c. Plastibell
d. Freehand cut sleeve resection

Answer: (c.) Plastibell. Devices for circumcision which leave behind an in situ ring, such as the Plastibell, carry the risk of the in situ ring being retained and causing postoperative edema and/or urinary obstruction. These devices are safe, and complications are rare, but the potential need for a provider to remove a retained ring means that these devices should only be used in settings where access to postoperative care is available.

Question 3: Which statement about circumcision is true?

a. Circumcision should always be performed to prevent penile cancer.
b. Circumcision should always be performed to prevent transmission of sexually transmitted diseases.
c. Circumcision should always be performed to prevent urinary tract infections.
d. It is safe and acceptable for parents to choose not to have their child circumcised.

Answer: (d.) It is safe and acceptable for parents to choose not to have their child circumcised. In many cultures, it is the norm for children not to be circumcised. From 1979 to 2010, the percentage of male newborns who underwent circumcision decreased from 65% to 58% [2]. Although some studies show a decrease in penile cancer, transmission of some sexually transmitted disease, and urinary tract infections, the reduction in risk is not sufficient to support uniform circumcision. Parents should be counseled that it is a perfectly normal choice not to choose circumcision for their child. However, the most compelling reason for circumcision is when parents choose it for cultural or religious reasons. The American Academy of Pediatrics statement from 2007 supports circumcision as an elective procedure for parents who choose it for their male children after proper counseling [1].

## References

1. Task Force on Circumcision. American academy of pediatrics. Male circumcision, technical report. Pediatrics 130(3): e756-e785, 2012. https://pediatrics.aappublications.org/content/130/3/e756. Accessed 22 Oct 2021.
2. Owings M, Uddin S, Williams S. Division of health care statistics, centers for disease control and prevention. Trends in circumcision for male Newborns in U.S. Hospitals: 1979–2010. https://www.cdc.gov/nchs/data/hestat/circumcision_2013/circumcision_2013.pdf. Accessed 22 Oct 2021.

# Hypospadias

**Jasmine I. Kashkoush and Robert Steckler**

**Key Points**

1) Hypospadias repair focuses on advancement of the urethra to create a normal passage of urine and semen in a cosmetically pleasing penis.
2) The diagnosis of a hypospadias made by physical exam may reveal a ventrally located urethral meatus, ventral penile curvature (chordee), and a dorsal hooded foreskin.
3) Do not continue with circumcision if you notice a hypospadias. Always refer to a surgical specialist.

## Hypospadias

1. Pathophysiology
    a. Results from arrested penile development
    b. Usually multifactorial
        i. Risk factors: premature birth, infants small for gestational age, intrauterine growth restriction, and assisted reproduction techniques
    c. Estimated to have incidence of 1/150–300 live male births
    d. Familial predominance with an index case resulting in incidence of NB 1/20 rate
    e. Associated anomalies

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    i. Cryptorchidism
    ii. Prostatic utricle
    iii. Penoscrotal transposition
    iv. Bifid scrotum
    v. Disorders of sexual development (DSD)
        1. Any child with hypospadias and UDTs, especially if dysmorphic or nonpalpable, should raise concern
        2. Most common is mixed gonadal dysgenesis, followed by varying degrees of androgen insensitivity

2. Diagnosis
    a. Diagnosed by physical exam
        i. Dorsal hooded foreskin
        ii. Ectopic, ventrally located urethral meatus
        iii. Ventral penile curvature
    b. There is a subset where foreskin is normal and abnormally large meatus only discovered when it is retracted—megameatus intact prepuce (MIP) varient
    c. Can also have a normally positioned meatus with ventral curvature (chordee)
    d. Meatus can be located along pathway of normal urethra distally from the glans and proximally to the perineum
    e. If there are many associated abnormalities, may be role for karyotyping, but it is not recommended in routine cases
3. Treatment
    a. General principles
        i. Over 200 repairs have been described
        ii. Suture material is surgeon's preference, as is use of post-op dressings and urinary diversion
        iii. Should be approached only if have undergone fellowship training
        iv. Use of optical magnification required
        v. All repairs should be followed through puberty
    b. Surgical repairs
        i. Most are performed in a single stage as outpatient
        ii. Very proximal lesions or those associated with severe curvature are commonly corrected in stages
        iii. All defects must be addressed including repositioning of meatus, ventral curvature, and scrotal abnormalities
        iv. Redo repairs and/or complex redo reconstructions are a special subset
            1. May require use of extragenital skin for urethroplasty.
            2. Most common is buccal or labial mucosa
                a. One- or two-staged approach

3. Severe shortage of shaft skin sometimes is the limiting factor
    a. Skin grafting
    b. Use of tissue expanders

c. Glanular hypospadias
    i. With or without curvature
    ii. Meatal advancement and glanuloplasty (MAGPI)—not uncommonly complicated by regression of the urethral meatus because of lack of ventral support for the advanced urethra
        1. M inverted V hypospadias repair is a better alternative in cases where the urethra is mobile and can be advanced to the tip of the glans
    iii. Division of web separating urethral pit from meatus
        1. Defect usually left open to re-epithelialize
    iv. Ventral curvature less than 30 degrees can generally be corrected with dorsal plication of the tunica albuginea of the corpora in the midline without corporotomy
        1. Avoid neurovascular bundle
    v. Use of urethral splints is based on surgeon's preference

d. Distal hypospadias
    i. Includes coronal and distal shaft lesions
    ii. Must be aware of dysplasia ventral shaft skin
    iii. Thiersch-Duplay repair—tubularization of the urethral plate with (tubularized incised plate—TIP) or without incision—is the most common technique employed
    iv. Results in a cosmetically and functionally superior glans and urethral meatus than other repairs
    v. Can be limited in children with a small glans
    vi. Very rarely the urethral plate in poorly developed and cannot be left intact
        1. Uncommonly may contribute to the ventral curvature
        2. May have to be divided
    vii. Coverage of the neourethra with a dartos fascial flap obtained from the dorsal prepuce is invaluable in reducing complications of a urethracutaneous fistula
    viii. Various repairs for ventral curvature
        1. Dorsal plication
        2. Mobilization of urethral plate

- ix. Use of urethral splints is based on the surgeon's preference
    - 1. Avoid use of balloon foley catheters
- x. Thiersch-Duplay or TIP repair can be extended to more proximal lesions such as midshaft and penoscrotal meatus

e. Proximal hypospadia

- i. Includes those at penoscrotal junction and perineum
- ii. May represent a variant of DSD
- iii. May be associated with a bifid scrotum, penoscrotal transposition, and ventral curvature
- iv. Commonly requires a staged approach (but single stage is an option used by some).
- v. Ventral curvature is corrected first
    - 1. The penis is degloved and curvature reassessed with an artificial erection
    - 2. Must be careful that dorsal plication will not reduce shaft length—to avoid shortening of the penis and to optimize straightening of the penis, mobilization or division of the urethral plate to allow placement of a ventral graft may be necessary
        - a. Dermal graft
        - b. SIS
- vi. Can then make decision to the tubularized urethral plate in a one-stage repair or approach in stages
    - 1. Must avoid any hair-baring skin
- vii. Prepuce is transposed ventrally
- viii. Delayed urethroplasty usually in 6–9 months
- ix. Dartos coverage of neourethra if possible or consider tunica vaginalis flap harvested from one of the testicles
- x. Urinary diversion with a urethral splint with or without a suprapubic cystotomy tube in toilet trained boys

4. Outcome

a. Complications

- i. Urethrocutaneous fistula (UCF)
- ii. Glans dehiscence
- iii. Persistent or recurrent curvature
- iv. Meatal stenosis
- v. Urethral stricture
- vi. Postoperative hematoma or wound infection

b. Redo repairs:

- i. Include most severe cases classified as a hypospadias cripple
- ii. Nature of repair can be complicated by the availability of shaft skin
    1. Creation of neourethra
    2. Shaft skin coverage
    3. Correction curvature
- iii. Can be simple UCF closure
    1. Avoid overlapping suture lines with the use of dartos interposition
    2. Operating microscope helpful
- iv. Redo Thiersch-Duplay repair can be used for most glans failures
- v. The most severe hypospadias complications may require skin grafting or buccal mucosa urethroplasty
    1. 1 or 2 stages
    2. Aid of plastic surgeon helpful
    3. Suprapubic tube diversion

## Additional Notes

The etiology of hypospadias is unclear but appears to be multifactorial. Associated risk factors include premature birth, infants small for gestational age, and intrauterine growth restriction. There appears to have an increased risk of hypospadias with patients who have undergone assisted reproductive births.

The gold standard of care is to repair a hypospadias with a surgical specialist. The timing of repair is important. It is important to balance the risks of surgery, anesthesia, and wound healing against the benefits of a successful repair. The general timing of repair based on the American Academy of Pediatrics recommends elective repair between 6 and 12 months of age.

## Study Questions

1) Question 1: A 3-month-old male is brought to the urology clinic because his mother notes that he has not had a normal stream. On exam, he has penile curvature, a ventrally located urethral meatus, and dorsal hood skin. What is his diagnosis?
    - a. Urethral stricture.
    - b. Hypospadias.
    - c. Meatal stenosis.
    - d. Redundant prepuce.

Answer to Question 1: (b.) Hypospadias. This boy likely has a hypospadias. Boys with hypospadias may have the triad of a penile curvature, a ventrally located urethral meatus, and dorsal hooded skin. More commonly, they present with a ventral urethral meatus and dorsal pit. Typically, a hypospadias is not harmful but may cause psychological distress for patients due to cosmetic reasons.

2) Question 2: A newborn is undergoing circumcision repair in the newborn nursery. Midway through the circumcision, the physician notices a hypospadias. What is the correct next step?

   a. Stop the procedure and refer to urology.
   b. Continue the circumcision.
   c. See the patient in 1 week to discuss circumcision.
   d. Proceed to the OR for hypospadias repair.

   Answer to Question 2: (a.) Stop the procedure and refer to urology. Patients with hypospadias should be referred to a surgical specialist for hypospadias repair. The recommended age for repair is between 6 and 12 months of age. The key is to never perform a circumcision in a patient with a hypospadias as there may not be enough skin left over during the hypospadias repair.

3) Question 3: A 12-month-old has just undergone hypospadias repair. What should he avoid doing in the first week after surgery?

   a. Avoid diaper changes.
   b. Playing on his tricycle.
   c. Using Tylenol.
   d. Applying warm compress to the wound.

   Answer to Question 3: (b.) Playing on his tricycle. Patients should avoid straddle toys like rocking horses and bicycles to help with his postoperative recovery. Patients may use over-the-counter pain medications and warm compresses as needed. There is no restriction to diaper changes.

## Further Reading

Partin AW, Dmochowski RR, Kavoussi LR, Peters CA, Wein AJ. Campbell Walsh Wein Urology. 12th ed. Amsterdam, NL: Elsevier; 2020.

Smith JA, Howards SS, Preminger GM, Dmochowski RR. Hinman's Atlas of Urologic Surgery Revised Reprint. 4th ed. Amsterdam, NL: Elsevier; 2019.

Kalfa N, Weiss DA, Hypospadias. AUA University. American Urological Association. Linthicum, MD, 2021, Online, https://university.auanet.org/core/pediatric/hypospadias/index.cfm, Accessed 6 Nov 2021.

# Pediatric Tracheostomy

**Vincent M. Desiato and Aileen P. Wertz**

**Key Concepts**

1) Multidisciplinary discussion with parents and extensive pre-operative counseling with the primary caregiver are paramount.
2) Medical optimization should be achieved prior to surgery, unless urgent/emergent intervention indicated.
3) In the operating room, patient positioned supine with neck extension. Attention to pediatric anatomical considerations: smaller tracheal diameter, a higher larynx, soft tracheal rings, and landmarks that are difficult to palpate.
4) Spare tracheostomy tubes (one of the same size and one a size smaller) should always be kept at bedside, as well as suction, suture removal kit, and lubrication. Items should travel with the patient during transport.
5) Pediatric tracheostomy tubes do not have inner cannulas, and are at higher risk of plugging or obstructing.
6) Patient safety and maintenance of the secure airway are of utmost importance, requiring careful attention and deliberate preparation, planning, and inter-departmental communication.

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1. *History*

   A. References to techniques similar to tracheostomy were first documented in 100 BC, but the earliest first-hand account was not until 1546, when Brasavola described using it to relieve airway obstruction caused by enlarged tonsils [1]. The term tracheostomy was derived from two Greek words meaning, "I cut the trachea," and was first used by Heister in 1739 [1, 2]. The procedure became more widespread in the early nineteenth century, used as a treatment of diphtheria. The technical aspects were later refined and described in detail by Chevalier Jackson in 1909 [2, 3]. More recent advancements include the emergence of positive-pressure ventilation and low-pressure cuffs on tracheostomy and endotracheal tubes, permitting widespread use in long-term ventilator support. Still, tracheostomy remains relatively uncommon in pediatric patients. In the USA, population-based incidence rates of tracheostomy decreased from 2003 to 2012, despite a statistically insignificant increase in the number performed from 2010 to 2018 [4, 5].

2. *Indications*

   A. The most common indications for tracheotomy in pediatric patients include prolonged mechanical ventilation, airway maintenance or pulmonary toilet, and bronchopulmonary dysplasia [6, 7]. One study reviewed indications for pediatric tracheotomy over the course of 30 years, and reported cardiopulmonary disease as the most common indication from 2010 to 2014 [8]. Additional common indications include hypotonia secondary to neurologic impairment, and traumatic injury [9, 10]. Prior to tracheotomy, a variety of interventions may serve as preventative or temporizing, and also ensure patient optimization prior to surgical intervention. Temporizing strategies may include nasopharyngeal airway, supplemental oxygen or heliox, racemic epinephrine, or antibiotics. In patients in whom endotracheal intubation is indicated, measures to avoid high cuff pressures on endotracheal tubes should be taken, and anti-reflux medications such as H2-antagonists or proton-pump inhibitors should be initiated. In select cases, patients with significant laryngomalacia may benefit from supraglottoplasty. In cases of subglottic stenosis, balloon dilation may be indicated prior to tracheotomy, and may avoid tracheotomy if adequate relief of airway obstruction is obtained.

      I. Upper airway obstruction

         i. Congenital

            a. Choanal atresia
            b. Macroglossia
            c. Laryngeal or subglottic stenosis
            d. Vocal fold paralysis

    ii. Infectious
        a. Epiglottitis
        b. Laryngotracheitis
        c. Ludwig’s angina
    iii. Severe sleep disorders
    iv. Trauma
        a. Facial or oral injury
        b. Burns/corrosives
        c. Laryngeal injury
    v. Neoplastic
  II. Respiratory insufficiency/ventilator dependence
    i. Congenital heart disease or heart failure
    ii. CNS disorders
    iii. Bronchopulmonary dysplasia
    iv. Prolonged intubation for other reasons
  III. Pulmonary toilet
    i. Recurrent aspiration or pneumonias
        a. Poor patient mucous clearance ability

3. *Patient assessment and pre-operative evaluation*
    A. Pre-operative counseling is paramount prior to performing a tracheotomy in a pediatric patient, and an in-depth discussion should take place. This should include multidisciplinary representation of neonatal or pediatric intensive care physicians, otolaryngologists, other consultants, as well as the parents or legal guardians of the patient. Caregivers should understand that tracheostomy will require frequent attention, and should be willing and able to provide the level of care necessary.
    B. Prior to surgery, the patient should be medically optimized. This includes low or stable ventilator requirements and normal coagulation parameters. If indicated urgently, optimization should focus on patient safety. Evaluation of the patient, including palpation of laryngotracheal landmarks, noting previous scars and cervicofacial anatomy is necessary in all cases. Special anatomical considerations in the pediatric patient include a smaller tracheal airway diameter and higher larynx in comparison to adults. Additionally, non-calcified tracheal rings are soft and can be difficult to palpate, along with other cervical landmarks.

4. *Preparation*
    - A. Anesthesia
        - I. General anesthesia is highly preferred
        - II. Establish secured airway
    - B. Patient positioning
        - I. Supine
        - II. Neck in extension with a shoulder roll in place
        - III. Tape in the submental area may be used for improved extension and exposure
    - C. Prepare and drape the patient
        - I. Face to be uncovered or accessible to anesthesia team for endotracheal tube removal
        - II. Palpate the patient's neck
        - III. Mark landmarks including thyroid and cricoid cartilages, suprasternal notch
        - IV. Mark planned incision—See below for location
        - V. Local anesthetic injected in the area of planned incision for hemostasis after type and dose discussed with anesthesia team
5. *Surgical procedure*
    - A. Incision
        - I. Skin incision one finger breadth above suprasternal notch and below the cricoid.
        - II. Incision may be horizontal or vertical, depending on surgeon's preference.
    - B. Dissection
        - I. Carry incision through the subcutaneous tissue.
            - i. May remove a small core of subcutaneous fat for improved exposure.
        - II. Dissection continued in the midline.
            - i. Frequent and repeated palpation of the trachea during dissection to ensure accuracy of approach and prevent deviation from midline.
        - III. Identification of anterior jugular veins and strap muscles
            - i. Anterior jugular veins retracted laterally or divided if identified.
            - ii. Strap muscles divided in the midline at midline raphe.
            - iii. Retract strap muscles laterally, using two Senn retractors, or other low-profile retractors.
        - IV. Address the thyroid gland
            - i. Retract or divide the thyroid isthmus.

        ii. May be divided with electro-cautery alone, or clamped on either side of the midline, divided and ligated with suture.

    V. Caution: airway fire

        i. When using high oxygen concentration, electro-cautery should be avoided after the airway has been entered or there is risk of the inadvertent airway entry, such as during thyroid division.

    VI. Stay sutures in trachea

        i. Placed bilaterally, prior to entering the airway.
        ii. Should be placed lateral to planned tracheal incision.
        iii. Vertically spanning two tracheal arches (ideally between 2 and 4).
        iv. Suture surgeon's choice. Authors prefer 2- or 3–0 monofilament.
        v. Sutures secured to chest and clearly labeled at the end of the procedure.

    VII. Tracheal incision

        i. Pre-oxygenation and communication with anesthesia team is critical at this point.
        ii. A vertical incision made in the midline of two tracheal rings

            a. Second and third, or third and fourth rings.
            b. In children, no cartilage or tissue is removed or excised.

    VIII. Placement of tracheostomy tube

        i. Gentle lateral traction placed on stay sutures to open stoma.
        ii. Orotracheal tube slowly pulled back under direct visualization.

            a. Draw back until existing orotracheal tube is *just above* tracheostomy.

        iii. Place the appropriate sized tracheostomy tube.
        iv. Confirm correct placement with flexible laryngoscope or bronchoscope.
        v. Secure tube with a snug tie around the neck (1 finger insertion with neck flexed) and four flange sutures (optional).
        vi. Once placement is confirmed and ventilation is satisfactory, orotracheal tube can be removed.

6. *Postoperative care*

    A. Chest x-ray immediately post-op

        I. Confirmation of tracheostomy tube position.
        II. Rule out pneumothorax secondary to procedure.

B. Intensive care setting immediately post-op (NICU, PICU)

  I. Sedation as needed based on patient age and health status to prevent tracheotomy tube dislodgement.

C. Pediatric tracheostomy tubes do not have inner cannulas, and are more susceptible to plugging or obstruction.

D. Humidified air

  I. Supplied via mechanical ventilator or tracheotomy collar.
  II. Gentle suctioning to the length of the tracheostomy tube.
  III. Instillation of saline prior to suctioning may assist in secretion management.

E. Management of cuff (if present)

  I. Cuff inflated if using pressure support.
  II. Cuff deflated if pressure support not necessary.

F. Neck skin care

  I. Keep skin clean and dry surrounding tracheostoma.

    i. Should be routine despite fresh tracheostomy. Will require multiple healthcare providers to perform safely.

  II. Look for and offload any pressure points where tracheostomy tube or ties are rubbing skin.

G. At the patient's bedside

  I. A spare tracheostomy tube of the same size and one size smaller should be kept with patient at all times.
  II. Suction: flexible suction catheters sized appropriately based on tracheotomy tube.
  III. Suture removal kit.
  IV. Lubrication.
  V. Items should travel with patient during transport.

H. First tracheostomy tube change

  I. Generally performed between five and seven days after the procedure.
  II. Stay sutures used for gentle retraction.
  III. Remove existing tube.
  IV. Place new tracheostomy tube.
  V. Confirm proper positioning with flexible fiberoptic endoscope.
  VI. Remove stay sutures.

7. *Potential complications*

A. Major complications

- Tube dislodgement

- False passage
- Mucous plugging
- Tracheal injury
- Tracheo-innominate fistula
- Pneumothorax
- Death

B. Minor complications

- Stoma breakdown
- Scarring
- Suprastomal granulations
- Intratracheal granulations/ulcerations

## Study Questions

1. The first tracheostomy tube change should generally be performed how many days after the procedure?

    a. 1 day
    b. 2–3 days
    c. 5–7 days
    d. 7–10 days
    e. 14 days or later

    Correct answer: (c.) 5–7 days.

**Explanation**
When the tracheostomy tract has epithelialized and the open edges of tracheal wall are securely healed to the peristomal tissues, the stoma is considered mature [11]. This concept forms the basis for planning the initial tracheostomy tube change. Maturation of the tracheostomy stoma generally occurs between postoperative days three and seven in children, but the exact timeline has not been established [12–14]. Initial tube changes are avoided for 48 to 72 hours after initial placement due to the risk of creating a false passage in an immature stoma [15–18]. To ensure the stoma is healed sufficiently, the surgical team typically replaces the initial tracheostomy tube after five to seven days in accord with expert panel consensus statements [14, 19]. A number of studies have investigated the role of early first tube changes, defined as prior to postoperative day five [12, 15, 17, 20, 21]. In the current literature, there is low-level evidence to support changing a tracheostomy tube on postoperative day three without the risk of increased complications, but no evidence supporting a role in decreasing hospital length of stay [18].

2. Electro-cautery should be avoided when using high oxygen concentration after the airway has been entered and when there is a risk of the airway inadvertently entered, such as during thyroid division due to risk of?

   a. Injury to recurrent laryngeal nerve
   b. Postoperative hypothyroidism
   c. Inadvertent esophageal injury
   d. Airway fire
   e. Intratracheal granulations

   Correct answer: (d.) Airway fire

**Explanation**

There are three fundamental components of fire, often referred to as the fire triad: an oxidizer, fuel, and a source of ignition.[22] Although fire triad components are present in the majority of surgical cases, all three must coincide to initiate fire. As a result, surgical fires most commonly occur during head and neck (28%) or airway (34%) surgery [23]. In a systematic review of 87 otolaryngic surgical fires, fire occurred during tracheostomy (36%) more often than any other surgical procedure [24]. Electro-cautery is the most common heat source, indicated in up to 90% of surgical fire incidents [24–26]. Endotracheal tubes are the most common source of fuel, and oxygen is the most common oxidizing agent involved [24]. Higher oxygen concentrations decrease the lower flammability limit of materials, meaning they ignite more easily and burn more rapidly [27]. Increased FiO2 is often required during tracheostomy, and 97% of surgical fires involved $FiO_2 > 30\%$ [24, 27]. Therefore, electro-cautery should be avoided after airway entry to prevent severe damage to the airway and lungs resultant from airway fire [28].

In the event of an airway fire, act quickly and follow a stepwise approach [29].

1. Immediate removal of the tracheal tube
2. Turn off any supplemental gases
3. Remove all flammable materials
4. Pour saline into the patient's airway
5. Reestablish ventilation by mask once fire is extinguished, avoiding supplemental oxygen if possible
6. Examine the tracheal tube integrity, due to concern for retained airway foreign body
7. Consider bronchoscopy

## References

1. Pierson DJ. Tracheostomy from A to Z: historical context and current challenges. Respir Care. 2005;
2. Frost EA. Tracing the tracheostomy. Ann Otol Rhinol Laryngol. 1976;85(5):618–24.
3. Witt CB Jr. The health and controversial death of George Washington. Ear Nose Throat J. 2001;80(2):102–5.

4. Friesen TL, Zamora SM, Rahmanian R, Bundogji N, Brigger MT. Predictors of pediatric tracheostomy outcomes in the United States. Otolaryngol Head Neck Surg. 2020;163(3):591–9.
5. Muller RG, Mamidala MP, Smith SH, Smith A, Sheyn A. Incidence, epidemiology, and outcomes of pediatric tracheostomy in the United States from 2000 to 2012. Otolaryngol Head Neck Surg. 2019;160(2):332–8.
6. Carron JD, Derkay CS, Strope GL, Nosonchuk JE, Darrow DH. Pediatric tracheotomies: changing indications and outcomes. Laryngoscope. 2000;110(7):1099–104.
7. Wetmore RF, Marsh RR, Thompson ME, Tom LW. Pediatric tracheostomy: a changing procedure? Ann Otol Rhinol Laryngol. 1999;108(7 Pt 1):695–9.
8. Gergin O, Adil EA, Kawai K, Watters K, Moritz E, Rahbar R. Indications of pediatric tracheostomy over the last 30 years: Has anything changed? Int J Pediatr Otorhinolaryngol. 2016;87:144–7.
9. Funamura JL, Durbin-Johnson B, Tollefson TT, Harrison J, Senders CW. Pediatric tracheotomy: indications and decannulation outcomes. Laryngoscope. 2014;124(8):1952–8.
10. Lawrason A, Kavanagh K. Pediatric tracheotomy: are the indications changing? Int J Pediatr Otorhinolaryngol. 2013;77(6):922–5.
11. Perkin RM, Newton DA, Swift JD. Pediatric hospital medicine: textbook of inpatient management. Lippincott Williams & Wilkins; 2008.
12. Deutsch ES. Early tracheostomy tube change in children. Arch Otolaryngol Head Neck Surg. 1998;124(11):1237–8.
13. Lewis CW, Carron JD, Perkins JA, Sie KC, Feudtner C. Tracheotomy in pediatric patients: a national perspective. Arch Otolaryngol Head Neck Surg. 2003;129(5):523–9.
14. Mitchell RB, Hussey HM, Setzen G, et al. Clinical consensus statement: tracheostomy care. Otolaryngol Head Neck Surg. 2013;148(1):6–20.
15. Lippert D, Hoffman MR, Dang P, McMurray JS, Heatley D, Kille T. Care of pediatric tracheostomy in the immediate postoperative period and timing of first tube change. Int J Pediatr Otorhinolaryngol. 2014;78(12):2281–5.
16. Othersen H. The Pediatric airway. edited by H. Biemann Othersen. *SERBIULA (sistema Librum 20).* 2021.
17. Van Buren NC, Narasimhan ER, Curtis JL, Muntz HR, Meier JD. Pediatric tracheostomy:timing of the first tube change. Otolaryngol Head Neck Surg. 2015;124(5):374–7.
18. Volsko TA, Parker SW, Deakins K, et al. AARC clinical practice guideline: management of pediatric patients with tracheostomy in the acute care setting. Respir Care. 2021;66(1):144–55.
19. Strychowsky JE, Albert D, Chan K, et al. International Pediatric Otolaryngology Group (IPOG) consensus recommendations: Routine peri-operative pediatric tracheotomy care. Int J Pediatr Otorhinolaryngol. 2016;86:250–5.
20. Chorney SR, Patel RC, Boyd AE, et al. Timing the first pediatric tracheostomy tube change: a randomized controlled trial. Otolaryngol Head Neck Surg. 2021;164(4):869–76.
21. Woods R, Geyer L, Mehanna R, Russell J. Pediatric tracheostomy first tube change: when is it safe? Int J Pediatr Otorhinolaryngol. 2019;120:78–81.
22. Yardley IE, Donaldson LJ. Surgical fires, a clear and present danger. Surgeon. 2010;8(2):87–92.
23. Pollock GS. Eliminating surgical fires: a team approach. AANA J. 2004;72(4):293–8.
24. Day AT, Rivera E, Farlow JL, Gourin CG, Nussenbaum B. Surgical fires in otolaryngology: a systematic and narrative review. Otolaryngol Head Neck Surg. 2018;158(4):598–616.
25. Kezze I, Zoremba N, Rossaint R, Rieg A, Coburn M, Schälte G. Risks and prevention of surgical fires: a systematic review. Anaesthesist. 2018;67(6):426–47.
26. Mehta SP, Bhananker SM, Posner KL, Domino KB. Operating room fires: a closed claims analysis. Anesthesiology. 2013;118(5):1133–9.
27. McManus N. Safety and health in confined spaces. Routledge; 2018.
28. Watson DS. Surgical fires: 100% preventable, still a problem. AORN J. 2009;90(4):589–93.
29. UbtCo S, Parameters P, Apfelbaum JL, et al. Practice advisory for the prevention and management of operating room fires: an updated report by the American Society of Anesthesiologists Task Force on Operating Room Fires. Anesthesiology. 2013;118(2):271–90.

# Part X
# Resources

# Coding for Pediatric Surgery

**Marc S. Lessin**

**Key Points**

1) Evaluation and management coding are an important aspect of pediatric surgery practice. There are unique codes specific for new and established office visits, inpatient services, ambulatory services, and consultation services.
2) Evaluation and management services generally cannot be billed in the postoperative period known as the “global period.” They are considered part of the surgical package. The duration of the global period is specific to the type of surgical procedure: specified as 0, 10, or 90 days. A single evaluation and management service can be billed prior to surgery. Each level of service is assigned a number of relative value units (RVUs) which are used to determine reimbursement.
3) Current Procedural Terminology (CPT) coding are 5-digit numeric codes used to report surgical services. Each code is associated with a number of relative value units which generate the amount which is billed/reimbursed. Several pediatric surgery index cases do not have specific codes and are reported using roughly similar codes. Sometimes unlisted codes for the organ system are reported for these cases.
4) A procedure done laparoscopically cannot be reported using the code for the open procedure if there is no specific laparoscopic code for that procedure. It is reported using an unlisted laparoscopic code for the surgical site or organ system.

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5) The Center for Medicare and Medicaid Services (CMS) publishes “correct coding edits” which designate procedure codes which cannot be reported together for the same operative session except under specific circumstances because these secondary codes are considered integral to the primary surgical procedure.
6) Modifiers are used to report services such as a preoperative service used to decide to perform surgery, when more than one procedure is performed in a single operative setting, assistant or co-surgeon services, and procedures performed on infants weighing less than 4 kg.

Coding is an important part of the practice environment. Billing and reimbursement is dependent on appropriate billing. Documentation is a key part of billing both for evaluation and management services and operative billing. An understanding of coding leads to prompt payment for services and avoids unnecessary delays and questioning of services. *Current Procedural Terminology* published yearly by the American Medical Association provides a list of 5-digit codes for evaluation and management services and surgical services. Reporting using these codes provides the basis for billing and reimbursement. Unfortunately, little is taught during surgical residency training about coding, billing, and how reimbursement works. This chapter provides an overview but not an exhaustive listing of codes and practices regarding how billing works relative to surgical services in the practice of pediatric surgery.

## Evaluation and Management Services

Billing begins with the initial encounter. Evaluation and management codes consist of a series of hierarchical codes depending on the following 3 categories: history, physical examination, and medical decision making. Each category involves specific bullet points which are necessary to achieve that level of service. The history consists of a chief complaint, history of the present illness, medications, allergies, social history, family history, and review of systems. The recognized review of systems categories consist of constitutional, eyes, ear, nose, and throat, cardiovascular, respiratory, gastrointestinal, genitourinary, musculoskeletal, integumentary, neurological, psychiatric, endocrine, hematologic/lymphatic, and allergic/immunologic. The physical examination consists of the following body areas: head, including the face, neck, chest including breasts and axilla, abdomen, genitalia, groin, buttocks back, and each extremity. The physical examination includes the following organ systems: eyes, ears, nose, mouth, and throat, cardiovascular, respiratory, gastrointestinal, genitourinary, musculoskeletal, skin, neurologic, psychiatric, and hematologic/lymphatic/immunologic. Electronic medical records often prepopulate the review of systems and physical examination findings which are then edited. The third element is medical decision-making which is straightforward, low complexity, moderate complexity, or high complexity. The series of codes depends on the setting in which they are rendered:

## Outpatient Services

- 99202–99205: Services rendered for a new patient which may be in the office setting or in the ER when the patient is not being admitted.
- 99211–99215: Office or other outpatient services for an established patient.

**Observation services** (Patient is being admitted but discharge is not expected beyond 2 past-midnight stays).

- 99218/ 99219 /99220: Observation level of care when admit and discharge are on different calendar dates.
- 99224/ 99225/99226: Subsequent observation care.
- 99234/ 99235/99236: Observation level of care when admit and discharge are on the same date of service.
- 99217: Observation care discharge services.

## Inpatient Services

- 99221 / 99222 / 99223: Initial hospital care for a new or established patient when the expected length of stay is greater than 2 post-midnight hospital days. Patient is admitted to a surgical service.
- 99231 / 99232 / 99233: Subsequent hospital care in the inpatient setting.
- 99238: Hospital discharge services from the inpatient level of care.
- 99251–99255: Inpatient consultation for a new or established patient for a patient cared for by another service.

Note that a single evaluation and management service can be reported prior to a surgical procedure. If the decision for surgery was made on that date of service, the evaluation and management code should be reported as a 5-digit code with the “-57” modifier which signifies decision for surgery (e.g., 99XXX.57). Postoperative services including daily care and discharge services should not be reported as they are considered part of routine postoperative care and are bundled into the typical postoperative follow-up care.

## Critical Care Billing

- 99291: Critical care, evaluation, and management for care of the critically ill or injured patient: first 30–74 minutes and 99292 for each additional 30 minutes. This code is used for evaluation of the severely injured trauma patient. The critically ill patient is defined as an illness or injury which acutely impairs one or more vital organ systems such that there is a high probability of imminent life-threatening deterioration in the patient's condition. This would be the appropriate code for a Level I trauma.

## Global Period/Reporting of Postoperative Services

The global period refers to the number of days for which routine postoperative care including daily inpatient services, discharge services, and outpatient follow-up is included in the surgical package and therefore is not reimbursed. In general, a single preoperative visit can be billed. If the decision for surgery was made based on that visit, the evaluation and management service should be reported with "-57" decision for surgery modifier. Surgical services are associated with 0, 10, and 90 day global periods depending on the surgical service. Most procedures for lesions of the integumentary system including incision and drainage of abscesses have a 10 day global period. Central venous catheter placement codes have a 10 day global period. Biopsy or excision of superficial lymph nodes has a 10 day global period; biopsy of deep axillary nodes has a 90 day global period. Most abdominal procedures including the common hernia repair, appendectomy, and cholecystectomy have a 90 day global period. Reporting of outpatient services within the global period can be reported with CPT code 99024—postoperative follow-up during the global period for a reason related to the original procedure.

## Surgical Services

Each surgical procedure is assigned a unique 5-digit CPT code. In some cases, there is no code which accurately reflects the procedure performed. In those cases, an unlisted code for the surgical site or organ system must be used. The American Medical Association guidelines state that when a procedure is performed laparoscopically and there is no laparoscopic specific code for that procedure, it is not appropriate to report the open code. In such instances, an unlisted laparoscopic code for the site or organ systems should be reported. If there is no laparoscopic code for that site or organ system (e.g., laparoscopic or thoracoscopic repair of diaphragm hernia), an unlisted [open] code for the site or organ system is reported. Each CPT code is assigned a number of relative value units. Reimbursement is determined by multiplying a dollar amount times the number of relative value units. Relative value units are determined based on the physician's work, practice expense, and professional liability issues. Adjustments are made for the setting in which care is delivered and regional differences in the cost of health care. The dollar multiplier is usually an amount determined by the Center for Medicare and Medicaid services or the insurance company. Fees for procedures reported under unlisted codes are determined by comparing fees for similar codes with adjustment up or down based on the degree of complexity of the procedure compared to the comparable code.

## Correct Coding Edits/Unbundling

Coding for surgical services requires that procedures that are considered an integral component of an operation are not separately reported. These procedures are considered "bundled." Reporting these codes is known as "unbundling." The Center for Medicare and Medicaid services (CMS) provides a two-column list of procedures by CPT code which cannot be reported together known as the National Correct Coding Initiative (NCCI). The Correct Coding Edit database for practitioners (PTP) can be queried from the government's public website (www.cms.gov) which lists codes which are not to be reported together. There are some exceptions which require an explanation. Each paired coding edit is accompanied by a modifier: "0" indicates that the coding pair should never be reported together on the same date of service; "1" indicated the codes may be reported together under certain circumstances but require a modifier and explanation. Examples of coding edits which do not allow procedures to be reported together would be an umbilical hernia during a laparoscopic procedure in which the hernia is used for port placement or excision of a spermatic cord lipoma in conjunction with inguinal hernia repair. Similarly, repair of a ventral hernia used for the opening of a laparotomy would be considered bundled. Closure including layered or undermining of a laparotomy incision cannot be reported separately unless closure requires a tissue rearrangement.

Some procedures are designated as "separate procedures" in the CPT booklet. These can never be reported in conjunction with another procedure. An example which makes little sense is a gastrostomy tube placed during a fundoplication. Gastrostomy tube placement is designated a "separate procedure." Correct coding edits do not allow reporting gastrostomy tube placement when performed in conjunction with open or laparoscopic fundoplication. Another example is lysis of adhesions whether done open or laparoscopically (44005 / 44180) which cannot be reported when another intra-abdominal procedure is performed. If an extensive and time-consuming lysis of adhesions is required, the primary procedure can be appended with the "-22" unusual procedural service modifier for additional reimbursement.

Laparoscopic operations sometimes require conversion to an open procedure. In these instances, only the open procedure is reported.

**Table 1** Common CPT modifiers. (Source: Marc Lessin/Adapted from Current Procedural Terminology, Professional Edition, American Medical Association, 2021)

| Modifier | Description |
|---|---|
| −22 | Unusual procedural service (increased complexity) |
| −24 | Unrelated evaluation and management (E & M) service by the same physical during the postoperative period |
| −25 | Significant, separately identifiable E & M service on the same day of procedure |
| −26 | Professional component (report with radiology services 76937 / 77001 |
| −47 | Anesthesia by surgeon |
| −50 | Bilateral procedure. Some insurance companies do not accept. Report as RT / LT or – 59 |
| −51 | Multiple procedures |
| −57 | Decision for surgery. Report when E & M service results in decision for surgery |
| −59 | Distinct procedural service |
| −62 | Two surgeons aka co-surgeons. Usually different specialties performing distinct parts of procedure |
| −63 | Procedure performed done infants less than 4 kg. Note several procedures already designated for neonates do not allow this modifier e.g. inguinal hernia repairs on infants <50 weeks post conceptual age. Include infant weight in operative report |
| −77 | Repeat procedure by another physician |
| −78 | Unplanned return to operating room by the same physician for related procedure during the postoperative period |
| −79 | Unplanned procedure or service by the same physician during the postoperative period |
| −80 | Assistant surgeon |
| −81 | Minimum assistant surgeon (non-physician assistant) |
| −82 | Assistant surgeon (when qualified resident surgeon not available at teaching hospital) |
| -RT | Right sided |
| -LT | Left sided |

## Modifiers

Modifiers are two-digit numeric or letter codes which are appended to five-digit CPT codes. They are important when reporting multiple procedures performed in the operative setting. A listing of common modifiers is attached (Table 1).

## Modifiers for Evaluation and Management Services

Modifier “-25”: Used when a patient presents on the day of a scheduled procedure and a significant separately identifiable service addressing a problem other than that for which the procedure is indicated.

Modifier “-57”: Decision for surgery. This modifier is used when the encounter involves the decision for surgery.

## CPT Modifiers for Surgical Services

The “-22” unusual procedure modifier is reported when the service involved is more than would be expected as described in the CPT code (e.g., an extensive lysis of adhesions required to perform the procedure). The “-26” modifier for professional component is reported in conjunction with ultrasound or fluoroscopy when used for central venous access. The code requires interpretation and documentation and should not involve the radiologist performing the same service. The “-51” modifier is used to reported multiple procedures. It should be reported when secondary procedures which are not integral to an operation are performed. These secondary procedures are generally subject to 50% fee reductions. Of note certain codes are deemed “add-ons” designated by a “+” sign in front of the code. These codes are exempt from the “-51” or “-59” modifier and are not subject to secondary procedure fee reductions. The “-59” is used when a secondary procedure is considered distinct from the primary procedure. It should be reported when there is a Correct Coding Edit which has a “1” attached meaning that the procedures may be reported together under certain circumstances such as a different anatomic site, organ system, incision/excision, separate lesion, or separate injury. The “-50” modifier is used for reporting bilateral procedures. Some insurance carriers including CMS do not allow reporting of this modifier but rather use a 2-line reporting using either the “-59” modifier or the RT/LT designation. The “-62” or co-surgeon modifier should be used when two surgeons, generally of different specialties, perform unique parts of an operation. Each should report their work separately using the same CPT code with the “-62” modifier added. Each should dictate a separate operative report describing their role. The “-63” modifier is an important modifier for pediatric surgeons. It is reported when a procedure is performed on a neonate weighing <4 kg. Of note, there are several procedures which are already unique to neonates which are exempt from the “-63” modifier (e.g., inguinal hernia repair in a premature infant < 50 weeks post-conceptual age or pyloromyotomy). (Tables 2 and 3).

**Table 2** CPT codes for common pediatric surgical procedures. (Source: Marc Lessin/Adapted from Current Procedural Terminology, Professional Edition, American Medical Association, 2021)

| CPT Code | Code explanation | Comment |
|---|---|---|
| | INTEGUMENTARY SYSTEM | |
| 10060 | Incision and drainage of abscess, simple | |
| 10061 | Incision and drainage of abscess, complicated or mutliple | |
| 11770 | Excision of pilonidal cyst or sinus; simple | |
| 11771 | Excision of pilonidal cyst or sinus; extensive | |
| 11772 | Excision of pilonidal cyst or sinus; complicated | |
| 19120 | Excision of cyst, fibroadenoma, or other benign tumor, breast | |

(continued)

**Table 2** (continued)

| **CPT Code** | **Code explanation** | **Comment** |
|---|---|---|
| 19300 | Mastectomy for gynecomastia | Report as bilateral (−50 modifier) or RT / LT |
| | MUSCULOSKELETAL/SOFT TISSUE | |
| 20680 | Removal of implant; deep | Code for removal of pectus bar |
| 20205 | Biopsy of muscle, deep | |
| 21011 | Excision tumor, soft itssue of face or scalp, subcutaneous <2 cm | E.g. excsion of dermoid |
| 21501 | Incision and drainage, deep abscess or hematoma, neck or thorax | |
| 21555 | Excsion, tumor soft tissue of neck or anterior thorax; < 3 cm | E.g. lipoma, dermoid excision |
| 21930 | Excision, tumor, soft itssue of back or flank, subcutaneous <3 cm | |
| 21931 | Excision, tumor, soft itssue of back or flank, subcutaneous ≥3 cm | |
| 21932 | Excision, tumor, soft tissue of back or flank, subfascial; <5 cm | |
| 21933 | Excision, tumor, soft tissue of back or flank, subfascial; ≥5 cm | |
| 22900 | Excision, tumor, soft tissue of abdominal wall, subfascial; <5 cm | |
| 22901 | Excision, tumor, soft tissue of abdominal wall, subfascial; ≥5 cm | |
| 27327 | Excision, tumor, soft tissue of thigh or knee; subcutaneous; <3 cm | |
| 27337 | Excision, tumor, soft tissue of thigh or knee; subcutaneous; ≥3 cm | |
| | RESPIRATORY | |
| 31600 | Tracheostomy, planned | |
| 31601 | Tracheostomy, younger than 2 years | |
| 31635 | Bronchoscopy with removal of foreign body | |
| 32607 | Thoracoscopy with diagnostic biopsy (e.g. wedge resection) | |
| 32650 | Thoracoscopy with pleurodesis | |
| 32653 | Thoracoscopy with removal of intrapleural foreign body or fibrin deposit | |
| 32655 | Thoracoscopy with resection of bullae including pleurodesis | |
| 32666 | Thoracoscopy with therapeutic wedge resection (mass or nodule), unilateral | |
| | VENOUS ACCESS | |
| 36555 | Insertion of non-tunneled centrally venous catheter; < 5 years of age | |
| 36556 | Insertion of non-tunneled centrally venous catheter; ≥ 5 years of age | |

**Table 2** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 36557 | Insertion of tunneled centrally venous catheter; < 5 years of age | |
| 36558 | Insertion of tunneled centrally venous catheter; ≥ 5 years of age | |
| 36560 | Insertion of tunneled centrally inserted port; < 5 years of age | |
| 36561 | Insertion of tunneled centrally inserted port; ≥ 5 years of age | |
| 76937 | Ultrasound guidance for central venous access | Add on code. Report with '-26' modifier |
| 77001 | Fluoroscopic guidance for central venous access | Add on code. Report with '-26' modifier |
| 36589 | Removal of tunneled central venous catheter; without port | E.g. removal of Broviac |
| 36590 | Removal of tunneled central venous catheter; with subcutaneous port | |
| | HEMATOPOIETIC | |
| 38100 | Splenectomy | "Separate procedure" |
| 38120 | Splenectomy, laparoscopic | |
| 38300 | Drainage of lymph node abscess, simple | |
| 38305 | Drainage of lymph node abscess, extensive | |
| 38500 | Biopsy or excision of lymph node(s); superficial | |
| 38510 | Biopsy or excsion of lymph node(s), deep cervical node(s) | |
| 38525 | Biopsy or excsion of lymph node(s), deep axillary node(s) | |
| 38531 | Biopsy or excsion of lymph node(s), deep inguinofemoral node(s) | |
| 38570 | Laparoscopic resection of retroperitoneal lymph node(s) | |
| | GASTROINTESTINAL | |
| 41010 | Incision of lingual frenulum (frenotomy) | |
| 43194 | Esophagoscpy, rigid with removal of foreign body | |
| 43215 | Esophagoscopy, flexible with removal of foreign body | |
| 43220 | Transendoscopic balloon dilation, esophagus | |
| 43246 | Esophagogastroduodenoscopy with directed placement of perc G-tube (PEG) | |
| 43280 | Laparoscopic fundoplication | |
| 43327 | Fundoplication; partial or complete; open | |
| 43450 | Dilation of esophagus by bougie | |
| 43453 | Dilation of esophagus over guidewire | |

(continued)

**Table 2** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 43500 | Gastrotomy with exploration or removal of foreign body | |
| 43653 | Laparoscopic gastrostomy | "Separate procedure:" cannot report with fundo |
| 43762 | Change of gastrostomy tube | Usually office of ER procedure |
| 43830 | Gastrostomy, open | "Separate procedure:" cannot report with fundo |
| 43831 | Gastrostomy, neonatal | "Separate procedure:" cannot report with fundo |
| 43840 | Gastrorrhaphy, suture of perforated ulcer, wound or injury | |
| 44005 | Enterolysis | "Separate procedure" |
| 44180 | Laparoscopic enterolysis | "Separate procedure" |
| 44602 | Enterorrhaphy for perforated ulcer, diverticulum, wound, injury or rupture | |
| 44604 | Colorrhaphy (large intestine) for perforation, without colostomy | |
| 44605 | Colorrhaphy (large intestine) for perforation, with colostomy | |
| 44615 | Intestinal stricturoplasy | |
| 44620 | Closure of enterostomy; large or small intestine | |
| 44625 | Closure of enterostomy; with resection and anastomosis other than rectal | |
| 44626 | Closure of enterostomy; with resection and colorectal anastomosis | |
| 44950 | Appendectomy; open | |
| 44955 | Appendectomy done for indicated purpose with another procedure | E.g. Ladd's procedure |
| 44960 | Appendectomy for ruptured appendicitis with abscess or peritonitis | |
| 44970 | Laparoscopic appendectomy | There is no laparoscopic code for ruptured appy |
| 45520 | Perirectal inection of sclerosing solution for prolapse | |
| 46020 | Placement of seton--rectal fistula | |
| 46040 | Incision and drainage of perirectal abscess | |
| 46045 | Incision and drainage of intramuscular abscess, transanal | |
| 46050 | Incision and drainage, perianal abscess, superifical | |
| 46060 | Incision and drainage of ischirectal or intramural abscess with fistulotomy | Placement of seton included when performed |
| 46270 | Surgical treatment of anal fistula; subcutaneous | |
| 46275 | Surgical treatment of anal fistula; intersphincteric | |
| 46280 | Surgical treatment of anal fistula; transphincteric, or multiple | Placement of seton included when performed |
| 46505 | Chemodenervation of interal anal sphincter | Botulinum toxin injection |

**Table 2** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 46608 | Anoscopy with remvoal of foreign body | |
| 49515 | Removal of fecal impaction or foreign body under anesthesia | |
| 47000 | Biopsy of liver, needle; percutaneous | |
| 47001 | Biopsy of liver, needle; when done in conjunction with another procedure | Add on code |
| 47100 | Biopsy of liver, wedge | |
| 47562 | Laparoscopic cholecystectomy | |
| 47563 | Laparoscopic cholecystectomy with cholangiogram | |
| 74300 | Cholangiography, intraoperative, radiological interpretation | Report with '-26' modifier if no radiologist |
| 47600 | Cholecystectomy; open | |
| 47605 | Cholecystectomy; open with cholangiogram | |
| 49000 | Exploratory laparotomy; open; with/without biopsy | Do not report if another procedure performed |
| 49002 | Reopening of recent laparotomy | |
| 49010 | Exploratory; retroperitoneal area with/without biopsy | |
| 49020 | Drainage of peritoneal abscess or localized peritonitis; other than appendix | Report for peritoneal drain for NEC w/ '-63' mod. |
| 49060 | Drainage of retroperitoneal abscess; open | |
| 49320 | Exploratory laparoscopy | Do not report if another procedure performed |
| 49321 | Exploratory laparoscopy with biopsy | |
| 49421 | Insertion of tunneled intraperitoneal catheter for dialysis | |
| 49422 | Removal of tunneled intraperitoneal dialysis catheter | |
| 49900 | Secondary repair of abdominal wall for evisceration or dehiscence | |
| 49905 | Omental flap, intra-abdominal | Add on code. Report in addition to primary proc. |
| | HERNIA CODES | |
| 49491 | Inguinal hernia preterm infant (< 37 weeks) up to 50 weeks post-conception | |
| 49492 | Premature infant inguinal hernia with incarceration or strangulation | |
| 49495 | Inguinal hernia repair full term infant <6 months or preterm infant >50 wks | |
| 49496 | Inguinal hernia repair full term infant <6 months with incarceration | |
| 49500 | Inguinal hernia repair; reducible 6 months to 5 years | |
| 49501 | Inguinal hernia repair; incarcerated 6 months to 5 years | |

(continued)

**Table 2** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 49505 | Inguinal hernia repair; >5 years; reducible | |
| 49507 | Inguinal hernia repair; >5 years; incarcerated ro strangulated | |
| 49520 | Repair recurrent inguinal hernia; any age; reducible | |
| 49521 | Repair recurrent inguinal hernia; any age; incarcerated or strangulated | |
| 49560 | Repair initial incisional or ventral hernia; reducible | |
| 49561 | Repair initial incisional or ventral hernia; incarcerated or stangulated | |
| 49565 | Repair recurrent incisional or ventral hernia; reducible | |
| 49566 | Repair recurrent incisional or ventral hernia; incarcerated or stangulated | |
| 49570 | Repair epigastric hernia; reducible | |
| 49572 | Repair epigastric hernia; incarcerated or strangulated | Report if incarcerated preperitoneal fat |
| 49580 | Repair umbilical hernia; <5 years; reducible | |
| 49585 | Repair umbilical hernia; >5 years; reducible | |
| 49650 | Laparoscopic inguinal hernia repair | Report with '-63' modifier if <4 kg |
| | UROGENITAL SYSTEM | |
| 51100 | Aspiration of bladder with insertion of suprapubic catheter | |
| 53060 | Drainage of Skene's gland abscess or cyst | |
| 54000 | Slitting of prepuce, dorsal or lateral; newborn | |
| 54001 | Slitting of prepuce, dorsal or lateral; except newborn | |
| 54150 | Circumcision, using clamp or other device with block | |
| 54160 | Circumcision, surgical excision other than clamp (neonate <28 days) | |
| 54160 | Circumcsion, surgical excision other than clamp (older than 28 days) | |
| 54162 | Lysis or excision of penile post-circumcision adhesions | |
| 54163 | Repair incomplete circumcision | |
| 54505 | Biopsy of testis, incisional | Separate procedure |
| 54600 | Reduction of torsion of testis, with or without fixation of opposite testicle | |
| 54620 | Fixation of contralateral testis | Separate procedure |
| 55000 | Puncture aspiration of hydrocele | |
| 55040 | Excison of hydrocele; unilateral | |
| 55041 | Excison of hydrocele; bilateral | |
| 55500 | Excison of hydrocele of psermatic cord, unilateral | |

**Table 2** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 55550 | Laparoscopy, surgical, with ligation of spermatic veins for varicocele | |
| 56405 | Incision and drainage of vulva or perineal abscess | |
| 56420 | Incision and drainage of Bartholin's gland abscess | |
| 56641 | Lysis of labial adhesions | |
| 56442 | Hymenotomy, simple incision | |

**Table 3** CPT codes for pediatric surgery index cases (Source: Marc Lessin/Adapted from Current Procedural Terminology, American Medical Association, 2021)

| CPT Code | Code explanation | Comment |
|---|---|---|
| | CHEST/LUNG/DIAPHRAGM | |
| 21601 | Excision of chest wall tumor including rib(s) | |
| 21743 | Reconstructive repair of pectus excavatum, (Nuss repair) w/ thoracoscopy | |
| 32480 | Lung; lobectomy | |
| 32482 | Lung; bilobectomy (2 lobes) | |
| 32662 | Thoracoscopy with removal of mediastinal cyst, tumor or mass | |
| 32663 | Thoracoscopy with lobectomy (single lobe) | |
| 33820 | Repair of patent ductus arteriosus; by ligation | Report this code for clipping of PDA |
| 33946 | ECMO cannulation; veno-venous | |
| 33947 | ECMO cannulation; arterio-venous | |
| 39200 | Resection mediastinal cyst | Bronchogenic cyst or esophageal duplication |
| 39220 | Resection mediastinal tumor | |
| 39503 | Repair congenital diaphragm hernia | |
| 39599 | Unlisted procedure: Thoracoscopic repair of congenital diaphragm hernia | There is no unlisted code for a thoracoscopic procedure |
| | GASTROINTESTINAL | |
| 43305 | Esophagoplasty with repair of tracheoesophageal fistula; cervical approach | H-type fistula |
| 43314 | Esophagoplasty with repair of tracheoesophageal fistula; thoracic approach | |
| 43520 | Pyloromyotomy; open | |
| 43659 | Unlisted procedure stomach reported for laparoscopic pyloromyotomy | No designated laparoscopic code |
| 43775 | Laparoscopic sleeve gastrectomy | |
| 44050 | Reduction of volvulus, intussusception, internal hernia by laparotomy | |

(continued)

**Table 3** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 44238 | Unlisted procedure, intestine for laparoscopic reduction inutussusception | No designated laparoscopic code |
| 44120 | Enterectomy, resection of small intestine, singel resection and anastomosis | |
| 44121 | Each additional resection and anastomosis with 44,120 | Add on code--exempt from muliple procedure modifier |
| 44126 | Enterectomy for congenital atresia, single resection and anastomosis | Code for small bowel atresia |
| 44127 | Enterectomy for congenital atresia, single resection with tapering | |
| 44128 | Each additional resection and anastomosis with 44,126 or 44,127 | |
| 44130 | Enteroenterostomy with or without cutaneous enterostomy | Repair duodenal atresia, duodenoduodenostomy |
| 44139 | Take-down of splenic flexure with parital colectomy | Add on code--exempt from muliple procedure modifier |
| 44140 | Colectomy, partial with anastomosis | |
| 44141 | Colectomy, partial with colostomy | |
| 44143 | Colectomy, partial with end colostomy and closure of distal segment | Hartmann procedure |
| 44144 | Colectomy, partial with resection with ostomy and mucous fistula | |
| 44145 | Colectomy, partial with resection with coloproctostomy | Low pelvic anastomosis |
| 44146 | Colectomy, partial with coloproctostomy and colostomy | |
| 44150 | Colectomy, total abdominal, without proctectomy with ileostomy | |
| 44155 | Colectomy, total abdominal with proctectomy with ileostomy | |
| 44157 | Colectomy, total abdominal with ileoanal anastomosis | |
| 44158 | Colectomy, total abdominal with ileal reservoir (J-pouch) | |
| 44160 | Colectomy, partial, with removal of terminla ileum with ileocolostomy | |
| 44320 | Colostomy or cecostomy | |
| 44340 | Colostomy revision, simple | |
| 44345 | Colostomy revision, complicated | |
| 44800 | Resection of Meckel's diverticulum or omphalomesenteric duct | |
| 44238 | Unlisted laparoscopic procedure, for laparoscopic approach | No designated laparoscopic code |
| 44820 | Excision of lesion of mesentery | Excision of intestinal duplication |

**Table 3** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 45100 | Biopsy of anorectal wall; anal approach | Suction or open rectal biopsy |
| 45120 | Proctectomy for congenital megacolon with pull through | Rectosigmoid segment Hirschsprung disease |
| 45121 | Proctectomy with subtotal or total colecotmy, with mulitple biopsies | Long segment Hirschsprung disease |
| 46716 | Repair of low imperforate anus with transposition of anoperineal fistula | |
| 46730 | Repair of high imperforate anus without fistula; perineal approach | |
| 46740 | Repair of high imperforate anus with rectovaginal fistula; perineal approach | |
| 46744 | Repair of cloacal anomaly; perineal approach | |
| 47010 | Hepatotomy, for open drainage of abscess or cyst | |
| 47120 | Hepatectomy; partial lobectomy | |
| 47122 | Hepatectomy; trisegmentectomy | |
| 47125 | Left hepatic lobectomy | |
| 47130 | Right hepatic lobectomy | |
| 47700 | Exploration for congenital atresia of the bile ducts with cholangiogram | |
| 47701 | Portoenterostomy (Kasai) | |
| 47715 | Excision of choledochal cyst | |
| 48140 | Pancreatectomy, distal subtotal, with or wihtout splenectomy | |
| 48146 | Pancreatectomy, distal, near total | |
| 48520 | Internal anastomosis of pancreatic cyst to gastrointestinal tract; direct | |
| 48547 | Duodenal exclusion with gastrojejunostomy for pancreatic injury | |
| 49203 | Excision / destruction intra-abdominal tumors; largest <5 cm | |
| 49204 | Excision / destruction intra-abdominal tumors; largest 5.1–10 cm | |
| 49205 | Excision / destruction intra-abdominal tumors; largest >10 cm | |
| 49600 | Repair of small omphalocel with primary closure | |
| 49605 | Repair of large omphalocele or gastroschisis with / without silo | |
| 49606 | Repair of large omphalocele or gastroschisis with removal of silo and closure | |
| | GENITOURINARY | |
| 50205 | Renal biopsy by surgical exposure of kidney | |
| 50220 | Nephrectomy, including partial ureterectomy, open approach | |

(continued)

**Table 3** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 50230 | Nephrectomy, radical w/ regional lymph node a/o cava thrombectomy | |
| 50240 | Nephrectomy, partial | |
| 50500 | Nephrorrhaphy, suture of kiden wound or injury | |
| 50545 | Laparoscopic radical nephrectomy | |
| 50546 | Laparoscopic nephrectomy | |
| 50548 | Laparoscopic nephrectomy with total uretectomy | |
| 51500 | Excison of urachal cyst or sinus, w/ wo umbilical hernia repair | |
| 51860 | Cystorrhaphy, suture of bladder wound, injury or rupture; simple | |
| 51865 | Cystorrhaphy, suture of bladder wound, injury or rupture; complicated | |
| 54520 | Orchiectomy; simple. Scrotal or inguinal approach | |
| 54530 | Orchiectomy, radical, for tumor; inguinal approach | |
| 54535 | Orchiectomy, radical, for tumor with abdominal exploration | |
| 54550 | Exploration for undescended testis (inguinal or scrotal area) | |
| 54560 | Exploration for undescended testis with abdominal exploration | |
| 54640 | Orchiopexy, inguinal or scrotal approach | |
| 54650 | Orchiopexy, abdominal approach for intra-abdominal testis | |
| 54690 | Laparoscopic orchiectomy | |
| 54692 | Laparoscopy, surgical; orchiopexy for intra-abdominal testis | |
| 58661 | Laparoscopy, surgical; with removal of adnexal structures (tube a/or ovary) | |
| 58700 | Salpingectomy, complete or partial, unilateral or bilateral | Separate procedure |
| 58720 | Salpingo-oophorectomy, complete or parital; unilateral or bilateral | Separate procedure |
| 58900 | Biopsy of ovary, unilateral or bilateral | Separate procedure |
| 58920 | Wedge resection or bisection of ovary; unilateral or bilateral | |
| 58925 | Ovarian cystectomy, unilateral or bilateral | |
| 58940 | Oophorectomy, partial or toal, unilateral or bilateral | |
| 58960 | Laparotomy for staging or restaging of ovarian, tubal or primary malignancy | |
| | ENDOCRINE SYSTEM | |
| 60220 | Total thyroid lobectomy, unilateral w/wo isthmusectomy | |

**Table 3** (continued)

| CPT Code | Code explanation | Comment |
|---|---|---|
| 60225 | Total thyroid lobectomy, unilateral with contrlateral subtotal lobectomy | |
| 60240 | Thyroidectomy, total or complete | |
| 60252 | Thyroidectomy, total or subtotal for malignancy with limited neck dissection | |
| 60254 | Thyroidectomy, total or subtotal for malignancy with radical neck dissection | |
| 60280 | Excision of thyroglossal duct cyst or sinus | |
| 60281 | Excision of thyroglossal duct cyst or sinus; recurrent | |
| 60540 | Adrenalectomy, partial or complete, or exploration of adrenal gland w/wo biopsy | |
| 60545 | Adrenalectomy with excsion of adjacent retroperitoneal tumor | |
| 60650 | Laparoscopic adrenalectomy | |

## Additional Notes

Coding is an essential component of the practice of surgery. A basic knowledge of coding is important for reimbursement, avoiding delays in payment, and proper reporting while avoiding improper reporting of both evaluation and management and surgical services. Coding begins with the initial assessment in the office or other settings such as the ER where evaluation and management services are reported. Generally, a single evaluation and management service during which the decision is made to perform surgery can be reported prior to the procedure. Updates of the history and physical prior to the surgical procedure cannot be billed. The coding for surgical procedures is based on 5-digit codes from *Current Procedural Terminology* (CPT codes). When more than one procedure is performed during a single operative setting, care must be taken to not bill procedures which are normally performed in conjunction with one another which is referred to as unbundling. This is accomplished by not reporting codes designated as “separate procedures” and checking correct coding edits to see if paired codes are disallowed. Note that services included in the “surgical package” include local anesthetic administration, immediate postoperative care, and typical postoperative follow-up care. Typical postoperative follow-up care during the global period cannot be billed but can be reported using CPT code 99024. The global period for typical postoperative care is 0 or 10 days for minor procedures and generally 90 days for major procedures.

The American College of Surgeons offers a free service to members to address coding questions. ACS Fellows are offered five free consultation services per calendar year for up to 10 minutes per consultation. Coding questions through this service are addressed to the ACS Coding Hotline website at *www.prsnetwork.com/acshotline*. A brief description of the question and any supporting documentation (with all identifying information removed) should be included. A member of the coding team will respond via e-mail.

## Study Questions

1) You perform a laparoscopic pyloromyotomy. You note there is no code for the procedure performed laparoscopically. Which of the following should be reported?

   a. Report using the code for pyloromyotomy 43520
   b. Report using the code for pyloromyotomy with the unusual procedural service modifier, i.e., 43520.22
   c. Report as unlisted procedure, stomach, i.e., 43999
   d. Report as unlisted laparoscopic procedure, stomach, i.e., 43659

   Answer to Question 1: D: Guidelines from the AMA state that when a procedure is performed laparoscopically, and there is no code for that procedure, an unlisted laparoscopic code should be reported. The code for the open procedure should not be reported.

2) Question 2: You perform a laparoscopic fundoplication and place a gastrostomy tube. Which of the following codes should be reported?

   a. 43280 for the fundoplication/43653.51 for laparoscopic gastrostomy tube
   b. 43280 for the fundoplication/43653.59 for laparoscopic gastrostomy tube
   c. Only report 43280
   d. Report 43280 for the fundoplication/43830 (open gastrostomy).

   Answer to Question 2: C: Gastrostomy tube placement is a “separate procedure.” It cannot be reported in conjunction with fundoplication whether done open or laparoscopically. Correct coding edits do not allow reporting of gastrostomy tube placement in conjunction with fundoplication.

3) You see a patient for uncomplicated appendicitis in the ER. Based on your evaluation, you decide to admit the patient to perform a laparoscopic appendectomy the following day. Regarding the evaluation and management service, which code should be reported?

   a. 99222.57: Admit to inpatient level of care with modifier for “decision for surgery.”
   b. 99219.57: Admit to observation level of care with modifier for “decision for surgery.”
   c. You cannot bill for the evaluation and management service.

   Answer to Question 3: B: Appendectomy for uncomplicated appendicitis is considered an outpatient procedure. Observation level of care is used when the hospital stay is expected to be less than 2 past-midnight days in the hospital. If the patient is to be operated upon and discharged the same day, the observation code 99234.57 should be reported. Normally, an evaluation and management (E/M) service cannot be reported on the day of a surgical procedure, but if the evaluation is the initial encounter, and the decision for surgery was made based on that encounter, the service can be reported with the "-57" modifier for decision for surgery.

## Further Reading

Current Procedural Terminology, Professional Edition, American Medical Association Pub. 2021.

Lieberman J, McNally M, Nagle J. Modifier 51 or 59? How to correctly report multiple procedures. Bull Am Coll Surg. 2019;14(4):50–1.

Ollapally V. Reporting global codes data in 2017. Bull Am Coll Surg. 2017;102(4):40–51.

National Correct Coding Edits, Center for Medicare and Medicaid Services (CMS); http://www.cms.hhs.gov/apps/ama/license.asp?file=/NationalCorrectCodInitEd

Smith S, McNally M, Lieberman J, Nagle J: 2021 CPT coding changes affect payment for E/M and other services, Bull Am Coll Surg, 2021; 106(1): 82–87.

# Note Templates for Inpatient Services

**Christopher P. Coppola**

1. Admission orders
    a. Admit
    b. Admitting service/physician/location /level of care (observation, intermediate, inpatient, ICU)
    c. Covering resident(s) and contact information
    d. Diagnosis
    e. Condition (stable, fair, poor, critical, moribund, etc.…)
    f. Vital sign frequency (also neurologic checks, pulse checks, weights, body part diameters, pulse oximetry)
    g. Continuous monitoring/telemetry needed
    h. Activity (bed rest, spine precautions, out of bed to chair, ambulate, assistance needed, fall risk, seizure prophylaxis)
    i. Allergies (medication, food, environmental, and reaction)
    j. Nursing procedures
        i. Wound and dressing care/changes
        ii. Parameters for which to call physician (temperature, pulse, blood pressure, respiratory rate, urine output, change in examination, function, or mental status)
        iii. Tubes and drains (both placement and monitoring of output)
        iv. Tubes/orifices which cannot be manipulated, any signs to post over bed
        v. Compression hose and/or sequential compression devices to prevent thrombosis
        vi. Glucose checks for diabetics

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   k. Diet or NPO orders
   l. In/out monitoring and frequency
   m. Intravenous access and fluids
   n. Medication

      i. Scheduled medication
      ii. Continuous infusions
      iii. Symptomatic medication (P.R.N. indications)
      iv. Deep venous thrombosis prophylaxis
      v. Stress ulcer prophylaxis
      vi. Home medications to stop/continue
      vii. Sliding scale for insulin

   o. Laboratory specimens to collect/process
   p. Blood bank orders for type and cross and/or transfusion
   q. Diagnostic imaging to obtain
   r. Electrocardiogram
   s. Services to consult
   t. Respiratory therapy orders
   u. Physical/occupational therapy orders

2. Preoperative note and checklist

   a. Diagnosis
   b. Planned procedure(s)
   c. Surgeon(s)
   d. Consent form status
   e. Anesthesia evaluation requested/accomplished
   f. Allergies
   g. Vital signs
   h. Relevant laboratory values

      i. Serum studies: Complete blood count, chemistries
      ii. Evaluation of coagulation
      iii. Urine
      iv. Blood bank type and cross or type and hold, how many units

   i. Relevant diagnostic imaging
   j. Electrocardiogram and other cardiac studies
   k. Evaluation of pulmonary function: pulmonary function test, arterial blood gas
   l. NPO order status and timing
   m. Intravenous fluids
   n. Preoperative antibiotics
   o. Bowel preparation
   p. Medications to continue/hold for operation

      i. Anticoagulation
      ii. Cardiac medication
      iii. Insulin

   q. Skin preparation/bathing

3. Brief operative note
    a. Patient name/record number
    b. Date
    c. Location
    d. Preoperative diagnosis
    e. Operation
    f. Postoperative diagnosis
    g. Surgeon
    h. Assistant(s)
    i. Anesthesia type
    j. Findings
    k. Fluids
        i. Intravenous fluid/blood volume administered
        ii. Estimated blood loss
        iii. Urine output
    l. Specimens for pathology and laboratory, intraoperative frozen section pathology report
    m. Drains and locations
    n. Diagram if necessary
    o. Condition
    p. Disposition location
    q. Physician responsible for operative report
4. Operative report
    a. Patient name/record number
    b. Date of operation and operative report
    c. Dictating physician
    d. Location
    e. Preoperative diagnosis
    f. Operation
    g. Postoperative diagnosis
    h. Attending surgeon and service
    i. Assistant(s)
    j. Findings
    k. Intravenous fluid/blood volume administered
    l. Estimated blood loss
    m. Urine output
    n. Specimens for pathology and laboratory, pathology results if available
    o. Intraoperative consultation
    p. Drains and tubes
    q. History and indications
    r. Consent obtained and documented

   s. Anesthetic used
   t. Antibiotics given
   u. Wound class
   v. Complications
   w. Deep venous thrombosis prophylaxis
   x. Positioning and padding
   y. Skin preparation
   z. Documentation of final time out
   aa. Narrative description of operation
   bb. Condition
   cc. Disposition location
   dd. Attestation of attending surgeon's presence and participation
   ee. Current Procedural Terminology (CPT) coding for procedure with applicable modifiers (see chapter on Coding in Pediatric Surgery)

5. Postoperative note
   a. Patient identification
   b. Time and location
   c. Diagnosis
   d. Operation performed
   e. Elapsed time since operation (4–6 hours are appropriate)
   f. Narration of events since operation
   g. Subjective account of patient's symptoms
   h. Control of pain
   i. Vital signs
   j. Intake of diet, tube feedings, and intravenous fluid
   k. Output of urine, tube drainage, vomiting, stool/stoma output
   l. Examination of wound and/or dressing, and drains
   m. Evaluation of distal neurovascular perfusion and function
   n. Postoperative lab results
   o. Overall assessment and plan

6. Progress note
   a. Patient identification
   b. Location
   c. Hospital day/postoperative day
   d. Diagnosis
   e. Operation
   f. Events of past 24 hours (interval history)
   g. Subjective account of patient's symptoms
   h. Vital signs
   i. Intake and output
   j. Diet
   k. Medications

        i. Continuous infusions
        ii. Antibiotic date, duration, indication for treatment
        iii. Symptomatic medication usage

    l. Stress ulcer prophylaxis
    m. Deep venous thrombosis prophylaxis
    n. Decubitus prophylaxis
    o. Head to toe examination
    p. Operative wound or dressing evaluation
    q. Evaluation of distal neurovascular perfusion and function
    r. Lab results
    s. Diagnostic imaging results
    t. Consultation reports
    u. Injury survey if trauma (tertiary survey)
    v. Assessment, system-based if critical care

        i. Neurologic
        ii. Respiratory/ventilator
        iii. Cardiac/circulation
        iv. Renal
        v. Fluids /electrolytes/nutrition
        vi. Gastrointestinal
        vii. Hematology
        viii. Immune/infectious disease
        ix. Endocrine
        x. Psychological/social
        xi. Extremities

    w. Plan

        i. Goals
        ii. Diet
        iii. Pain control
        iv. Antibiotics
        v. Consultations

    x. Discharge planning and estimated date of discharge

7. Trauma tertiary survey

    a. Date and time of trauma tertiary survey
    b. Date and time of admission
    c. Patient identification
    d. Location
    e. Mechanism of injury
    f. Known injuries
    g. Past medical history

        i. Illnesses
        ii. Operations
        iii. Medications
        iv. Allergies
        v. Habits (ethanol, tobacco, others)

    h. Physical examination
    i. Laboratory values
    j. Urinalysis
    k. Substance abuse screening

        i. Ethanol
        ii. Drugs of abuse

    l. Clinical/trauma management guidelines applicable
    m. Radiographic studies and results
    n. Cervical spine clearance
    o. DVT prophylaxis
    p. Stress ulcer prophylaxis
    q. Medication reconciliation
    r. Pending diagnostic tests
    s. Consultation results
    t. Consultations pending
    u. Incidental findings

8. Discharge summary

    a. Patient identification
    b. Physician completing summary
    c. Location
    d. Type of admission (observation/ inpatient/intermediate care/ICU)
    e. Admission date
    f. Discharge date
    g. Diagnosis, primary and secondary
    h. Discharging service
    i. Discharging physician
    j. Disposition destination
    k. Condition on discharge
    l. Diet
    m. Activity
    n. Medications on discharge
    o. Future appointments
    p. Pending test results
    q. Future testing
    r. Brief admission history and physical

    i. Complaint
    ii. History of present illness
    iii. Past medical/surgical history
    iv. Mediations/allergies
    v. Social history and family history
    vi. Birth history and vaccinations for infants

s. Hospital course
t. Procedures performed
u. Complications
v. Laboratory test results
w. Diagnostic imaging results
x. Discharge plan
y. Providers who should receive discharge summary

# Common Medications

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The authors are not responsible for product liability, negligence, injury, or otherwise for any loss, harm, or injury resulting from any material contained in this work. This work contains information relating to general principles of medical care which should not be construed as specific instructions for individual patients. The responsibility for making treatment decisions lies with the providers caring for specific patients. Pharmaceutical product inserts and manufacturers' product information should be reviewed for current information, including contraindications, dosages, and precautions. Users of this text accept these conditions.

1. Acetaminophen (Tylenol): Non-narcotic analgesic, antipyretic (Table 1).
   - Oral:
     - Infants, children, adolescents: 10–15 mg/kg/dose every 4–6 h as needed; do not exceed 5 doses in 24 h (max dose 75 mg/kg/day not to exceed 4 g/day).
     - Children ≥12 years, Adult: 325–650 mg every 4–6 h or 1 g 3–4 times/day (max dose 4 g/day).
   - Rectal:
     - Infants, children <12 years: 10–20 mg/kg/dose every 4–6 h as needed; do not exceed 5 doses in 24 h (max dose 75 mg/kg/day not to exceed 4 g/day).
   - IV:
     - Infants and children <2 years: 15 mg/kg/dose every 6 h (max 60 mg/kg/day).

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**Table 1** Classes of medications covered in this chapter

| **Analgesics** | **Anti-microbials** | | **GI** |
|---|---|---|---|
| • Acetaminophen (Tylenol)<br>• Aspirin<br>• Buprenorphine (Subutex)<br>• Fentanyl<br>• Gabapentin (Neurontin)<br>• Hydromorphone (Dilaudid)<br>• Liposomal Buprenorphine (Exparel)<br>• Ibuprofen (Motrin)<br>• Methadone<br>• Morphine | • Amoxicillin and Clavulanate (Augmentin)<br>• Ampicillin<br>• Ampicillin Sulbactam (Unasyn)<br>• Cefazolin (Ancef)<br>• Cefepime (Maxipime)<br>• Cefoxitin (Mefoxin)<br>• Ceftriaxone (Rocephin)<br>• Cephalexin (Keflex)<br>• Ciprofloxacin<br>• Clindamycin | • Ertapenem (Invanz)<br>• Fluconazole (Diflucan)<br>• Gentamicin<br>• Ketoconazole<br>• Meropenem (Merrem)<br>• Metronidazole (Flagyl)<br>• Nitrofurantoin (Macrobid)<br>• Piperacillin-tazobactam (Zosyn)<br>• Trimethoprim-sulfamethoxazole (Bactrim)<br>• Vancomycin | • Bisacodyl (Dulcolax)<br>• Cholestyramine (Prevalite)<br>• Docusate (Colace)<br>• Erythromycin<br>• Lactulose<br>• Lansoprazole (Prevacid)<br>• Omeprazole (Prilosec)<br>• Polyethylene glycol (Miralax) |
| **Neuro/Sedatives** | **Coagulants/ Anti-coagulants** | **Miscellaneous** | **Electrolyte Repletion** |
| • Dexmedetomidine (Precedex)<br>• Diphenhydramine (Benadryl)<br>• Flumazenil (Romazicon)<br>• Ketamine<br>• Levetiracetam (Keppra)<br>• Lorazepam (Ativan)<br>• Midazolam (Versed)<br>• Phenobarbital<br>• Phenytoin (Dilantin) | • Enoxaparin (Lovenox)<br>• Heparin<br>• NovoSeven<br>• Tranexamic Acid | • Albuterol<br>• Crofab<br>• Furosemide (Lasix)<br>• Heparin-saline (line flushes) | • Calcium<br>• Sodium<br>• Magnesium<br>• Potassium<br>• Phosphate |

    - Children ≥2 years, < 50 kg: 15 mg/kg/dose every 6 h or 12.5 mg/kg/dose every 4 h (max single dose 750 mg, max daily dose 75 mg/kg not to exceed 3750 mg).
    - Children ≥2 years, ≥ 50 kg: 15 mg/kg/dose every 6 h or 12.5 mg/kg/dose every 4 h (max single dose 1000 mg, max daily dose 75 mg/kg not to exceed 4 g).

  - Considerations: avoid chronic use in hepatic impairment.

2. Albuterol (AccuNeb, ProAir HFA, Proventil HFA, Ventolin HFA, VoSpire ER): Beta2-Adrenergic agonist agent, bronchodilator.

   - Acute exacerbation, asthma:

     - Infants, children ≤5 years:

       - Inhalation: 90 mcg/spray, 2–6 puffs every 20 min for 2–3 doses, then every 1–4 h as needed.
       - Nebulization: 2.5 mg every 20 min for the first hour, then every 1–4 h as needed.

- Children ≥6 years:
    - Inhalation: 90 mcg/spray, 2–10 puffs every 20 min for 2–3 doses, then every 1–4 h.
    - Nebulization: 2.5–5 mg (every 20 min for 3 doses then 2.5–10 mg every 1–4 h as needed or 10–15 mg/h by continuous nebulization.
- Adult:
    - Inhalation: 90 mcg/spray, 2–4 puffs every 20 min for 3 doses, then every 1–4 h.
    - Nebulization: 2.5–5 mg every 20 min for 3 doses, then every 1–4 h as needed or 10–15 mg continuous nebulization over 1 h.

3. Amoxicillin and clavulanate potassium (Augmentin): Antibiotic, Beta-lactam and Beta-lactamase combination, Penicillin.
    - Pediatric general dosing (Oral):
        - 4:1 formulation: 20–40 mg amoxicillin/kg/day divided doses 3 times daily (max 1500 mg/day)
        - 7:1 formulation: 25–45 mg amoxicillin/kg/day divided doses 2 times daily (max 1750 mg/day)
        - 14:1 formulation: 90 mg amoxicillin/kg/day divided doses 2 times daily (max 4000 mg/day)
        - 16:1 extended-release formulation: children ≥40 kg
            - 2000 mg amoxicillin every 12 h
    - Adult general dosing:
        - Oral: 500 mg every 8–12 h or 875 mg every 12 h; extended release 2 g every 12 h.
        - IV: 1 g every 8 h or 2 g every 8–12 h.
4. Ampicillin: penicillin antibiotic.
    - Pediatric:
        - IV/IM: 50–200 mg/kg/day divided every 6 h (max 8 g/day).
        - Oral: 50–100 mg/kg/day divided every 6 h (max 2000 mg/day).
    - Adult:
        - IV/IM: 2 g every 4 h.
        - Oral: 500 mg every 6 h.
5. Ampicillin Sulbactam (Unasyn): Beta-lactam and Beta-lactamase combination, antibiotic, penicillin.
    - General dosing:
        - Pediatric:

    - IV/IM: 100–200 mg ampicillin/kg/day divided every 6 h (max 2000 mg/dose).
  - Adult:
    - IV/IM: 1.5–3 g every 6 h (max 12 g/day).
- Surgical Prophylaxis (IV): given 60 min prior to incision, repeat dose every 2 h.
  - Pediatric: 75 mg/kg (max 3 g).
  - Adult: 3 g.

6. Aspirin: Analgesic, antipyretic, Nonsteroidal Anti-Inflammatory Drug, Salicylate.
   - Typically, aspirin is avoided in children less than 12 years who actively exhibit flu symptoms or are recovering from chickenpox due to association with Reye's syndrome.
   - Analgesia:
     - Infants, children, adolescents < 50 kg (oral, rectal): 10–15 mg/kg/dose every 4–6 h (max daily dose 90 mg/kg not to exceed 4 g/day).
     - Children ≥12 years ≥50 kg (oral, rectal): 325–650 mg every 4–6 h (max 4 g/day).
     - Adult (oral, rectal): 325 mg to 1 g every 4–6 h (max 4 g/day).
7. Bisacodyl (Dulcolax): Laxative, stimulant.
   - Pediatric:
     - Oral:
       - ≥3 years to <10 years: 5 mg once daily
       - 10–12 years: 5–10 mg once daily
       - ≥12 years: 5–15 mg once daily
     - Suppository, enema:
       - 2–10 years: 5 mg once daily
       - >10 years: 5–10 mg once daily
   - Adult:
     - Oral: 5–15 mg once daily
     - Suppository, enema: 10 mg once daily
8. Buprenorphine (Subutex): Opioid analgesic, opioid partial agonist.
   - Acute Pain (moderate to severe):
     - Children 2–12 years (opioid naïve):
       - IM, slow IV: 2–6 mcg/kg/dose every 4–6 h.
     - Adolescents and Adult (opioid naïve):

- IM or slow IV: 0.3 mg every 6–8 h as needed, can give second dose within 30–60 min if necessary.

- Opioid Dependence: *combination sublingual tablet with naloxone* (Oral).
  - Induction: 2–4 mg, monitor for precipitated withdrawal for 60–90 min, if tolerated, can increase in 2–4 mg increments, titrate to dose that is clinically effective for 24 h.
  - Maintenance: daily doses are often ≥8 mg, maximum dose 24 mg/day.

9. Cefazolin (Ancef): First-generation cephalosporin antibiotic.
   - General dosing:
     - Pediatric:
       - IV/IM: 25–150 mg/kg/day divided every 8 h (max 6 g/day).
     - Adult:
       - IV/IM: 1–2 g every 8 h.
   - Surgical Prophylaxis (IV/IM): given 60 min prior to incision, repeat dose every 4 h.
     - Pediatric:
       - Neonatal: 25 mg/kg (repeat dosing every 6–8 h).
       - Infants, children, adolescents: 30 mg/kg (max 2 g).
     - Adult:
       - < 120 kg: 2 g
       - ≥ 120 kg: 3 g
10. Cefepime (Maxipime): Fourth generation cephalosporin antibiotic.
    - General dosing:
      - Pediatric (IV/IM):
        - Non-*pseudomonas*: 50 mg/kg/dose every 12 h (max 2 g/dose).
        - *Pseudomonas*: 50 mg/kg/dose every 8 h (max 2 g/dose).
      - Adult (IV):
        - Non-*pseudomonas*: 1–2 g every 8–12 h.
        - *Pseudomonas*: 2 g every 8 h.
    - Surgical prophylaxis (IV): given 60 min prior to incision, repeat dose every 4 h.
      - Pediatric:
        - Neonatal: 30 mg/kg (no repeat dosing).
        - Infants, children, adolescents: 50 mg/kg (max 2 g).

    - Adult: 2 g.
  - Considerations: consider dosing adjustments with impaired kidney function.

11. Cefoxitin (Mefoxin): Second-generation cephalosporin antibiotic.
    - General dosing:
      - Pediatric (IV/IM):
        - >3 months: 80–160 mg/kg/day divided doses every 4–6 h (max 12 g/day)
      - Adult (IV/IM):
        - 1–2 g every 6–8 h (max 12 g/day)
    - Surgical Prophylaxis: given 60 min prior to incision.
      - Pediatric (IV/IM):
        - Neonatal: 33 mg/kg (no repeat dosing).
        - Infants, children, adolescents: 40 mg/kg (max 2 g, repeat every 4 h).
      - Adult (IV/IM):
        - 2 g (repeat every 2 h)
12. Ceftriaxone (Rocephin): Third generation cephalosporin antibiotic.
    - General dosing:
      - Pediatric (IV/IM):
        - Mild to moderate infection: 50–75 mg/kg/dose once daily (max dose 1000 mg).
        - Severe infection: 100 mg/kg/day divided every 12–24 h.
      - Adult (IV/IM):
        - 1–2 g once daily
    - Surgical prophylaxis: given 60 min prior to incision.
      - Pediatric (IV): 50 mg/kg (max 2 g).
      - Adult (IV): 2 g.
13. Cephalexin (Keflex): First-generation cephalosporin antibiotic.
    - Pediatric (oral):
      - Mild to moderate infection: 25–50 mg/kg/day divided every 6–12 h (max 2 g/day).
      - Severe infection: 75–100 mg/kg/day divided every 6–8 h (max 4 g/day).

- Adult (oral):
  - 250 mg to 1 g every 6 h or 500 mg every 12 h (max 4 g/day).

14. Cholestyramine (Prevalite, Questran): Antilipemic agent, bile acid sequestrant.
    - Children ≤10 years (Oral): 240 mg/kg/day in 2–3 divided doses (max 4 g/day).
    - Children ≥10 years (Oral): 240 mg/kg/day before breakfast (max 8 g/day).
    - Adult: 4 g once or twice daily, titrate up as needed (max 16 g/day in 2 divided doses).
15. Ciprofloxacin (Cipro): Quinolone antibiotic.
    - Pediatric:
      - IV: 10 mg/kg/dose every 8–12 h (max 400 mg/dose).
      - Oral: 10–20 mg/kg/dose twice daily (max 750 mg/dose).
    - Adult:
      - IV: 400 mg every 12 h.
      - Oral: 500 mg every 12 h.
16. Clindamycin (Cleocin): Antibiotic, anaerobic.
    - Pediatric:
      - Oral:
        - Mild to moderate infections: 10–25 mg/kg/day divided every 6–8 h (max 1800 mg/day).
        - Severe infections: 30–40 mg/kg/day divided every 6–8 h (max 1800 mg/day).
      - IV/IM:
        - Mild to moderate infections: 20 mg/kg/day divided every 8 h (max 1800 mg/day).
        - Severe infections: 40 mg/kg/day divided every 6–8 h (max 2700 mg/day).
    - Adult:
      - Oral: 600–1800 mg/day in 2–4 divided doses (max 2400 mg/day in 4 divided doses).
      - IV/IM: 600–2700 mg/day in 2–4 divided doses (max 4800 mg/day IV in 4 divided doses; max 600 mg/dose IM).
17. Crofab (Ovine polyvalent Crotalidae North and South American snake antivenom): Antivenin.
    - Children ≥1 year, Adult (IV):
      - Initial dose: 4–6 vials within 6 h of snakebite, repeat with additional 4–6 vials if control not achieved, maximum initial dose 12 vials.

    - 8–12 vials initially in patients with immediate life-threatening effects
  - Maintenance dose: 2 vials every 6 h for up to 18 h.

18. Dexmedetomidine (Precedex): Alpha-adrenergic agonist agent, sedative.
    - Pediatric:
      - Loading dose (IV): 0.5–1 mcg/kg over 10 min (optional).
      - Maintenance (IV): 0.2–0.5 mcg/kg/h, children < 1 year may require higher infusion rates (max 1 mcg/kg/h, or 2.5 mcg/kg/h in intubated patients).
    - Adult:
      - Loading dose (IV): 1 mcg/kg over 10 min (optional).
      - Maintenance (IV): 0.2–1 mcg/kg/h, infusion rates of 1.5–2.5 mcg/kg/h can be used in intubated patients.
19. Diphenhydramine (Benadryl): antihistamine, sedative.
    - Pediatric (Oral/IV/IM):
      - 2 to <6 years: 6.25 mg every 4–6 h (max 37.5 mg/day)
      - 6 to <12 years: 12.5–25 mg every 4–6 h (max 150 mg/day).
      - ≥ 12 years: 25–50 mg every 4–6 h (max 300 mg/day)
    - Adult:
      - Oral: 25 mg every 4–6 h or 50 mg every 6–8 h.
      - IV/IM: 10–50 mg every 6 h (max 400 mg/day).
20. Docusate (Colace): laxative, surfactant, stool softener.
    - Pediatric:
      - Oral (Docusate sodium):
        - 2 to <12 years: 50–150 mg/day in single or divided doses
        - ≥12 years: 50 to 360 mg/day in single or divided doses
      - Oral (Docusate calcium):
        - ≥12 years: 240 mg once daily
      - Rectal:
        - 2 to <12 years: 100–283 mg once daily
        - ≥12 years: 283 mg 1–3 times daily
    - Adult:
      - Oral:
        - Docusate sodium: 50–360 mg daily or in divided doses.
        - Docusate calcium: 240 mg once daily.

- Rectal:
  - 283 mg 1–3 times daily

21. Enoxaparin (Lovenox): Anticoagulant, low molecular weight heparin.
    - Thrombosis prophylaxis (SC):
      - Neonatal:
        - 0.75 mg/kg/dose every 12 h
      - Infants ≥2 months, children:
        - 0.5 mg/kg/dose every 12 h
      - Adult:
        - 40 mg daily
    - Thrombosis treatment (SC):
      - Neonatal:
        - Initial: 1.5 mg/kg/dose every 12 h.
        - Maintenance: titrate 4-to-6-h post-dose anti-factor Xa level between 0.5–1 units/mL.
      - Infants ≥2 months, children:
        - Initial: 1 mg/kg/dose every 12 h.
        - Maintenance: titrate 4-to-6-h post-dose anti-factor Xa level between 0.5–1 units/mL.
      - Adult:
        - 1 mg/kg every 12 h or 1.5 mg/kg once daily
    - Considerations: monitor anti-Xa levels closely in patients with renal impairment, may require dosage adjustment or discontinuation of therapy, discontinue if platelets < 100,000.
22. Ertapenem (Invanz): Carbapenem antibiotic.
    - Pediatric:
      - Infants, children (IV/IM): 15 mg/kg/dose twice daily (max 500 mg/dose).
      - Adolescents (IV/IM): 1000 mg once daily.
    - Adult:
      - IV/IM: 1 g daily.
23. Erythromycin ethylsuccinate (E.E.S, Eryped): Macrolide antibiotic, prokinetic agent.
    - GI motility agent dosing:

    - Pediatric:
        - Oral: 3 mg/kg/dose 4 times daily (max 10 mg/kg or 250 mg).
    - Adult:
        - IV: 3 mg/kg/dose administered over 45 min every 8 h.
        - Oral: 250–500 mg 3 times daily.

24. Fentanyl: Narcotic analgesic.
    - Pediatric (IV):
        - Acute pain:
            - Infants: 1–2 mcg/kg/dose every 2–4 h (younger infants may require up to 4 mcg/kg/dose).
            - Children: 1–2 mcg/kg/dose every 30–60 min.
            - Adolescents <50 kg: 0.5–1 mcg/kg/dose every 1–2 h.
            - Adolescents ≥50 kg: 25–50 mcg every 1–2 h.
        - Continuous analgesia/sedation:
            - Infants and children: 1–2 mcg/kg bolus, then continuous infusion at 1–3 mcg/kg/h.
            - Adolescents <50 kg: 0.5–1 mcg/kg bolus, then continuous infusion at 0.5–2 mcg/kg/h.
            - Adolescents ≥50 kg: 25–50 mcg bolus, then continuous infusion at 25–200 mcg/h.
    - Adult (IV):
        - Severe pain (IV/IM):
            - 50–100 mcg every 30–60 min
        - Continuous analgesia/sedation (IV):
            - 1–2 mcg/kg or 25–100 mcg, then 25–200 mcg/h

25. Fluconazole (Diflucan): Antifungal agent.
    - Pediatric (IV, Oral):
        - 6–12 mg/kg on day 1, then 3–12 mg/kg once daily
    - Adult (IV, Oral):
        - Fixed dosing: 800 mg on day 1, then 400 mg once daily.
        - Weight-based dosing: 12 mg/kg on day 1, then 6 mg/kg once daily.

26. Flumazenil (Romazicon): benzodiazepine antidote.
    - Pediatric (IV):
        - Benzodiazepine reversal for sedation/anesthesia:

        - Initial dose 0.01 mg/kg (max 0.2 mg) given over 15 seconds, can repeat dose after 45 seconds, then every minute as needed (max cumulative dose 0.05 mg/kg or 1 mg, whichever is lower).
    - Benzodiazepine overdose:
        - Bolus dosing: Initial dose 0.01 mg/kg (max 0.2 mg) can repeat dose every minute as needed (max cumulative dose 1 mg).
        - Continuous infusion dosing: Initial dose 0.01 mg/kg (max 0.2 mg) followed by continuous infusion of 0.005–0.01 mg/kg/h.
- Adult (IV):
    - Benzodiazepine reversal for sedation/anesthesia:
        - Initial dose 0.2 mg over 15 seconds, can repeat dose every 1 min as needed (max 5 doses or 1 mg).
    - Benzodiazepine overdose:
        - Initial dose 0.2 mg over 30 seconds, then 0.3 mg over 30 seconds if needed, can repeat 0.5 mg doses over 30 seconds every 1 min as needed (max cumulative dose 3 mg).

27. Gabapentin (Neurontin): Anticonvulsant, analgesic, GABA analog.
    - Pediatric (Oral):
        - Seizure adjunctive therapy:
            - Children 3–12 years:
                - Initial dose: 10–15 mg/kg/day divided every 8 h, titrate over ~3 days.
                - Maintenance dose: 25–40 mg/kg/day divided every 8 h.
            - Children >12 years:
                - Initial dose: 300 mg 3 times daily.
                - Maintenance dose: 900–1800 mg/day 3 times daily (max 3600 mg/day).
        - Neuropathic Pain:
            - Titrating schedule:
                - Day 1: 5 mg/kg at bedtime (max 300 mg/dose).
                - Day 2: 5 mg/kg/dose twice daily.
                - Day 3: 5 mg/kg/dose 3 times daily (max daily dose 3600 mg/day).
    - Adult (Oral):
        - Seizure adjunctive therapy:
            - 300 mg 3 times daily, titrate as needed

- Neuropathic pain:
  - 100–300 mg 1–3 times daily, titrate as needed

28. Gentamicin: Aminoglycoside antibiotic.
   - Pediatric:
     - Conventional dosing (IV/IM): 2–2.5 mg/kg/dose every 8 h.
     - Weight-based extended interval dosing (IV): 5–7.5 mg/kg/dose every 24 h.
     - Age based extended interval dosing (IV):
       - ≥3 months to <2 years: 9.5 mg/kg/dose every 24 h
       - 2 to <8 years: 8.5 mg/kg/dose every 24 h
       - ≥8 years: 7 mg/kg/dose every 24 h
   - Adult:
     - IV/IM: 3–5 mg/kg/day in divided doses every 8 h.
   - Surgical prophylaxis: given 60 min prior to incision.
     - Pediatric (IV):
       - Neonatal: 4 mg/kg.
       - Infants, children, adolescents: 2.5 mg/kg (max 400 mg).
     - Adult (IV):
       - 5 mg/kg
29. Heparin (unfractionated): Anticoagulant.
   - Thrombosis treatment (IV):
     - Neonatal:
       - Loading dose: 75 units/kg IV over 10 min.
       - Maintenance dose: 28 units/kg/h continuous IV infusion, titrate to maintain anti-Xa level 0.35–0.7 units/mL.
     - Children and adolescents:
       - Loading dose: 75 units/kg IV over 10 min.
       - Maintenance dose: 20 units/kg/h continuous IV infusion, titrate to maintain anti-Xa level 0.35–0.7 units/mL.
     - Adult:
       - Loading dose: 80 units/kg IV bolus, or 5000 unit bolus.
       - Maintenance dose: 18 units/kg/h continuous IV infusion.
   - Thrombosis prophylaxis (SC):

    - Adult:
        - 5000 units every 8–12 h

30. Heparin-Saline for line flushing.
    - Notes
        - 2–5 mL/flush depending on catheter size.
        - Dose of heparin flush used should not approach therapeutic unit per kg dose.
        - Heparin flush should be given if stagnant blood is observed in catheter, after used for drug or blood administration, or after blood withdrawal from catheter.
    - Pediatric:
        - <10 kg: 10 units/mL concentration
        - >10 kg: 100 units/mL concentration
    - Adult:
        - 100 units/mL concentration
31. Hydromorphone (Dilaudid): Narcotic analgesic, antitussive.
    - Pediatric:
        - Infants <6 months weighing >10 kg:
            - Oral: 0.03–0.06 mg/kg/dose every 4 h.
            - IV: 0.01–0.015 mg/kg/dose every 3–6 h.
            - Continuous IV: 0.003–0.005 mg/kg/h.
        - <50 kg:
            - Oral: 0.03–0.08 mg/kg/dose every 3–4 h.
            - IV: 0.015 mg/kg/dose every 3–6 h.
            - Continuous IV: 0.003–0.005 mg/kg/h.
        - ≥50 kg:
            - Oral: 1–2 mg every 3–4 h.
            - IV: 0.2–0.6 every 2–4 h.
            - Continuous IV: 0.3 mg/h.
            - IM, SC: 0.8–1 mg every 4–6 h.
            - Rectal: 1–2 mg every 3–6 h.
    - Pediatric PCA:
        - ≥5 years weighing <50 kg:
            - Usual concentration: 0.2 mg/mL.
            - Demand dose: 0.003–0.005 mg/kg.

    - Lockout: 5 doses/h.
    - Lockout intverval: 6–10 min.
    - Usual basal rate: 0–0.004 mg/kg/h.
  - ≥50 kg:
    - Usual concentration: 0.2 mg/mL.
    - Demand dose: 0.05–0.4 mg.
    - Lockout interval: 5–10 min.
- Adult:
  - Oral: 2–4 mg every 4–6 h.
  - IV: 0.2–1 mg every 2–3 h.
  - Continuous IV: 0.5–3 mg/h.
  - IM, SC: 1–2 mg every 2–3 h.
  - Rectal: 3 mg every 6–8 h.
- Adult PCA:
  - Loading dose: 0.4 mg.
  - Demand dose: 0.1–0.4 mg.
  - Lockout interval: 10 min.

32. Ibuprofen (Motrin, Advil): Non-narcotic analgesic, Nonsteroidal Anti-Inflammatory drug.
    - Pediatric (Oral):
      - ≥6 months: 4–10 mg/kg/dose every 6–8 h (max 400 mg/dose, max 40 mg/kg/day or 3200 mg/day, whichever is less)
    - Adult (Oral):
      - 200–400 mg every 4–6 h or 600–800 mg every 6–8 h (max 3.2 g/day)
33. Ketamine (Ketalar): General anesthetic, noncompetitive NMDA receptor antagonist.
    - Pediatric:
      - Sedation/analgesia, procedural:
        - ≥3 months:
          - IM: 4–5 mg/kg as single dose, can repeat dose after 5–10 min.
          - IV: 1–2 mg/kg over 30–60 seconds, can repeat 0.5–1 mg/kg doses every 5–15 min.
          - Intranasal: 3–6 mg/kg (half dose per nostril).
          - Oral: 5 mg/kg (with Midazolam) given 30–45 min before procedure.
          - Rectal: 1.5–3 mg/kg (with Midazolam) 20 min prior to procedure.
      - Sedation, critically ill patients (IV):

    - ≥5 months:
      - Initial dose 0.5–2 mg/kg, then 5–20 mcg/kg/min (0.3–1.2 mg/kg/h) continuous IV infusion.
  - Adult:
    - Procedural sedation:
      - IV: 1–2 mg/kg over 1–2 min, can repeat dose 0.5–1 mg/kg every 5–10 min.
      - IM: 4–5 mg/kg, can repeat 2–5 mg/kg dose after 5–10 min.
    - Sedation, critically ill patients, intubated (IV):
      - 0.1–0.5 mg/kg bolus, then 0.2–0.5 mg/kg/h continuous IV infusion

34. Ketoconazole: Antifungal agent.
    - Pediatric (Oral):
      - ≥2 years: 3.3–6.6 mg/kg/day once daily (max 400 mg/day)
    - Adult:
      - 200 mg/day once daily, can increase to 400 mg once daily if needed
35. Lactulose: Ammonium detoxicant, laxative.
    - Constipation:
      - Pediatric (Oral):
        - 1–2 g/kg/day divided once or twice daily
      - Adult (Oral):
        - 10–20 g daily, may increase to 40 g daily if needed
36. Lansoprazole (Prevacid): Gastric acid secretion inhibitor, proton pump inhibitor.
    - Pediatric (Oral):
      - Infants: 2 mg/kg/day, divided once or twice daily.
      - Children and adolescents: 0.7–3 mg/kg/day, divided once or twice daily (max 30 mg/day).
    - Adult (Oral):
      - 15–30 mg once daily for up to 8 weeks
37. Levetiracetam (Keppra): Anticonvulsant.
    - Seizure prophylaxis, Traumatic Brain Injury (TBI):
      - Pediatric (IV, Oral):
        - Limited data available, optimal dose not defined.
          5–55 mg/kg/day (median 20 mg/kg/day) divided twice daily

    - Maximum Doses:
        - 20–40 kg: 1500 mg/day
        - >40 kg: 3000 mg/day
  - Adult (IV):
    - Loading dose: 20 mg/kg (round to nearest 250 mg) infused over 5–15 min.
    - Maintenance dose: 1 g infused over 15 min every 12 h for 7 days (max dose 1.5 g every 12 h).

38. Liposomal Bupivacaine [marcaine] (Exparel): Local anesthetic.
    - Children ≥6 years (local infiltration):
      - 4 mg/kg once, maximum dose 266 mg/dose (20 mL)
    - Adult:
      - Single dose, dose dependent on size of site, maximum dose 266 mg/dose (20 mL).
39. Lorazepam (Ativan): Benzodiazepine.
    - Pediatric:
      - Anxiety:
        - Infants, children <12 years (Oral, IV): 0.05 mg/kg/dose every 4–8 h (max 2 mg/dose).
        - ≥12 years (Oral): 0.25–2 mg/dose 2–3 times daily (max 2 mg/dose)
      - Sedation:
        - Oral: 0.05 mg/kg.
      - Status epilepticus:
        - IV: 0.1 mg/kg (max 4 mg/dose) slow IV, can repeat dose once in 5–10 min.
    - Adult:
      - Anxiety:
        - IV/IM/Oral: 0.5–2 mg every 4–6 h (max 10 mg/day).
      - Sedation:
        - IV: 1–4 mg, then 1–4 mg every 2–6 h (max 4 mg single dose).
      - Status epilepticus:
        - IV: 4 mg at a maximum rate of 2 mg/min, can repeat at 3–5 min if seizure persists.

40. Methadone: Opioid analgesic.
    - Pain (severe):
      - Infants ≤6 months, nonventilated:
        - IV, SC: 0.025 mg/kg/dose every 4–8 h.
        - Oral: 0.025 to 0.05 mg/kg/dose every 4–8 h.
      - Infants >6 months, children, nonventilated:
        - IV, SC:
          - <50 kg: 0.1 mg/kg/dose every 4–8 h
          - ≥50 kg: 5–8 mg every 4–8 h
        - Oral.
          - <50 kg: 0.1–0.2 mg/kg/dose every 4–8 h
          - ≥50 kg: 5–10 mg every 4–8 h
      - Adult:
        - IM, IV, SC: 2.5–10 mg every 8–12 h, can increase dose by 2.5 mg every 5–7 days.
        - Oral: 2.5–5 mg every 8–12 h, can increase dose by 2.5 mg every 5–7 days.
    - Iatrogenic opioid dependency:
      - Prevention: consider in patients receiving opioids >14 days, transition to methadone, once stabilized, wean 10–20% of original dose every 24 to 48 h.
      - Treatment:
        - Pediatric (Oral, IV):
          - Initial: 0.05–0.1 mg/kg/dose every 6 h, increase by 0.05 mg/kg/dose until withdrawal symptoms controlled.
          - Maintenance: lengthen dosing interval to 12–24 h.
          - Discontinuation: wean as tolerated until dose is 0.05 mg/kg/day, then discontinue.
        - Adult (Oral, IV):
          - Initial: titrate to ~40 mg/day in divided doses to stabilize.
          - Maintenance: continue 40 mg/day dose for 2–3 days.
          - Discontinuation: decrease daily dose by 20% every 1–2 days, keep dosing sufficient to treat withdrawal symptoms.
    - Considerations: long half-life, may accumulate and cause sedation; may require reduction in dose with renal impairment.

41. Meropenem (Merrem): Carbapenem antibiotic.
    - Pediatric:
        - IV: 20 mg/kg every 8 h (max 1000 mg/dose).
    - Adult:
        - IV: 500 mg over 30 min every 6 h, or 1–2 g every 8 h.
42. Metronidazole (Flagyl): Amebicide, antiprotozoal, anaerobic antibiotic.
    - Pediatric general dosing:
        - Oral: 15–50 mg/kg/day divided 3 times daily (max 2250 mg/day).
        - IV: 22.5–40 mg/kg/day divided 3–4 times daily (max 4000 mg/day).
    - Adult general dosing:
        - Oral/IV: 500 mg every 8 h.
    - Surgical prophylaxis: given 60 min prior to incision.
        - Pediatric (IV):
            - Neonatal: 15 mg/kg (repeat dose every 6–8 h).
            - Infants, children, adolescents: 15 mg/kg (max 500 mg).
        - Adult:
            - IV: 500 mg.
43. Midazolam (Versed): Benzodiazepine.
    - Sedation/amnesia:
        - Pediatric:
            - IM: 0.1–0.15 mg/kg 30–60 min before procedure (max dose 10 mg).
            - Oral: >6 months: 0.25–0.5 mg/kg once (max dose 20 mg).
            - IV:
                - ≥6 months to <6 years: Initial 0.05–0.1 mg/kg carefully titrated, (max dose 6 mg)
                - ≥6 years: Initial 0.025–0.05 mg/kg carefully titrated (max dose 10 mg)
            - Adolescents: 1–2.5 mg over 2+ min, carefully titrated (max dose 10 mg).
            - Rectal: >6 months: 0.25–0.5 mg/kg once (max dose 20 mg).
            - Intranasal: 0.2–0.8 mg/kg single dose, can repeat dose in 15 min (max dose 10 mg).
        - Adult:

- IV: 0.5–2 mg over 2+ min, repeat every 2–5 min as needed (max total dose 5 mg).
- Intranasal (using injectable solution): 0.1 mg/kg 15 min prior to procedure (max dose 10 mg).

44. Morphine: Opioid analgesic.

- Pediatric:
    - Infants ≤6 months:
        - Nonventilated:
            - Oral: 0.08–0.1 mg/kg/dose every 3–4 h.
            - IV, SC: 0.025–0.03 mg/kg/dose every 2–4 h.
        - Ventilated:
            - IV: 0.05 mg/kg/dose every 2–4 h.
            - Continuous IV: 8–20 mcg/kg/h.
    - Infants >6 months, children, adolescents:
        - Oral:
            - <50 kg: 0.2 = 0.5 mg/kg/dose every 3–4 h (max 15–20 mg)
            - ≥50 kg: 15–20 mg every 3–4 h
        - IV/IM/SC:
            - <50 kg: 0.05 mg/kg/dose every 2–4 h
            - ≥50 kg: 2–5 mg every 2–4 h
        - Continuous IV/SC infusion:
            - <50 kg: 0.01 mg/kg/h
            - ≥50 kg: 1.5 mg/h
    - PCA, children ≥5 years <50 kg:
        - Usual concentration 1 mg/mL.
        - Demand dose 0.01–0.03 mg/kg/dose.
        - Lockout 5 doses/h.
        - Lockout interval: 6–8 min.
        - Usual basal rate 0–0.03 mg/kg/h.
    - PCA, children ≥5 years ≥50 kg:
        - Usual concentration 1 mg/mL.
        - Demand dose: 0.5–2.5 mg.
- Adult:
    - Oral: 10 mg every 4 h (max 30 mg every 4 h for severe pain).
    - IV: 1–4 mg every 1–4 h (max 10 mg every 4 h for severe pain).

    - Rectal: 10 mg every 4 h.
    - PCA, IV:
        - Usual concentration: 1 mg/mL.
        - Demand dose: 0.5–2 mg.
        - Lockout interval: 5–10 min.
        - Max cumulative dose: 7.5 mg within 1 h.

45. Nitrofurantoin (Macrobid, Macrodantin, Furadantin): antibiotic.
    - Pediatric:
        - UTI:
            - Oral: 5–7 mg/kg/day divided every 6 h (max 100 mg/dose).
        - UTI prophylaxis:
            - Oral: 1–2 mg/kg/day single dose at bedtime or divided twice daily (max 100 mg/day).
    - Adult (Oral):
        - UTI:
            - Nitrofurantoin monohydrate/macrocrystals (Macrobid): 100 mg twice daily.
            - Nitrofurantoin macrocrystals (Furadantin, Macrodantin): 50–100 mg every 6 h.
        - UTI prophylaxis:
            - Nitrofurantoin monohydrate/macrocrystals (Macrobid): 100 mg once daily at bedtime.
            - Nitrofurantoin macrocrystals (Furadantin, Macrodantin): 50–100 mg once daily at bedtime.
46. NovoSeven RT (Coagulation Factor VIIa {recombinant}): Antihemophilic agent.
    - Off-label use for fulminant hepatic failure, liver transplantation, drug-induced coagulopathy, platelet disorders, renal failure, ICH, post-operative or intraoperative hemorrhage, bleeding due to trauma, and post cardiopulmonary bypass coagulopathy.
        - Neonatal and Pediatric (IV):
            - 90 mcg/kg once, can repeat dose in 2-h intervals as needed
                - *No maximum dose established*
        - Adult (IV):
            - 90 mcg/kg once, can repeat dose in 2-h intervals as needed
47. Omeprazole (Prilosec): Gastric acid secretion inhibitor, proton pump inhibitor.

- Infants and children (Oral):
  - 5 to <10 kg: 5 mg once daily
  - 10 to <20 kg: 10 mg once daily
  - ≥20 kg: 20 mg once daily
- Adult (Oral):
  - 10–40 mg once daily

48. Phenobarbital: Barbiturate, anticonvulsant.
    - Pediatric:
      - Seizures:
        - Infants, children ≤5 years: 3–5 mg/kg/day in 1–2 divided doses.
        - >5 years: 2–3 mg/kg/day in 1–2 divided doses
      - Status epilepticus (IV):
        - 15–20 mg/kg (max 1000 mg/dose), can repeat with 5–10 mg/kg dose in 10 min
    - Adult:
      - Seizures:
        - Oral, IV: 2 mg/kg/day in divided doses.
      - Status epilepticus (IV):
        - 15–20 mg/kg single dose (infused at 50–100 mg/min), can repeat with 5–10 mg/kg after 10 min
49. Phenytoin (Dilantin): Anticonvulsant.
    - Seizure prophylaxis, Traumatic Brain Injury (TBI):
      - Pediatric (IV):
        - Loading dose: 18–20 mg/kg over 20 min.
        - Maintenance: 6 mg/kg/day divided every 8 h.
      - Adult (IV, Oral):
        - Loading dose: 17–20 mg/kg (infused at ≤50 mg/min, max dose 2 g).
        - Maintenance:
          - Begin 8–12 h after loading dose.
          - 100 mg every 8 h or 5 mg/kg/day (round to nearest 100 mg) every 8 h
50. Piperacillin-tazobactam (Zosyn): Antibiotic, extended penicillin, Beta-lactam and Beta-lactamase.
    - General dosing (IV):

    - Pediatric:
        - Infants 2–9 months: 270 mg/kg/day divided every 8 h (max 16 g/day).
        - >9 months <40 kg: 337.5 mg/kg/day divided every 8 h
        - >9 months: >40 kg: 3.375 g every 6 h
    - Adult:
        - 3.375 g IV every 6 h
  - Surgical prophylaxis (IV): given 60 min prior to incision, repeat dose every 4 h.
    - Pediatric:
        - 85 mg/kg/dose (max 3.375 g)
    - Adult:
        - 3.375 g

51. Polyethylene glycol (Miralax, Glycolax): Laxative, osmotic.
    - Pediatric (Oral):
        - Constipation:
            - 0.2–0.8 g/kg/day (max 17 g/day)
        - Bowel preparation (children > 2 years):
            - 1.5 g/kg/day for 4 days (max 100 g/day)
    - Adult (Oral):
        - Constipation:
            - 17 g once daily
        - Bowel preparation:
            - 17 g in 8 oz. of clear liquid every 10 min until 2 L are consumed (used in conjunction with bisacodyl or magnesium citrate)
52. Tranexamic Acid (Cyklokapron): Antifibrinolytic, antihemophilic, hemostatic agent.
    - Prevention of perioperative bleeding:
        - Neonatal and Pediatric general dosing (IV):
            - Loading dose: 10–30 mg/kg.
            - Maintenance dose: 5–10 mg/kg/h continuous IV, target serum concentration of 20–70 mcg/ml.
        - Adult general dosing (IV):

    - 1 g (or 10–30 mg/kg) over 10–30 min prior to procedure, followed by continuous infusion intraoperatively or repeat post-op bolus
  - Acute traumatic coagulopathy (hemorrhagic):
    - Children <12 years (IV):
      - Loading dose: 15 mg/kg over 10 min within 3 h of injury (maximum dose 1000 mg/dose).
      - Maintenance dose: 2 mg/kg/h for ≥8 h or until bleeding stops.
    - Children >12 years and Adult (IV):
      - Loading dose: 1000 mg over 10 min within 3 h of injury.
      - Maintenance dose: 1000 mg infused over 8 h.
  - Diffuse alveolar hemorrhage (intractable):
    - Children ≤25 kg (IV):
      - 250 mg/dose every 6 h for 3–4 doses
    - Children >25 kg and adolescents (IV):
      - 500 mg/dose inhaled every 6 h for 3–4 doses
    - Treatment Response:
      - Response (+): repeat for 2–3 more doses.
      - Response (−): add inhaled recombinant factor VIIa (3-day maximum duration of therapy).
  - Considerations: may require dosage adjustment with kidney impairment.

53. Trimethoprim-sulfamethoxazole (Bactrim): Sulfonamide antibiotic.
    - Pediatric (Oral, IV):]
      - ≥2 months: 6–12 mg/kg/day divided every 12 h (max 160 mg/dose)
    - Adult:
      - Oral: 1–2 double strength tablets (160 mg TMP/ 800 mg SMX) every 12–24 h.
      - IV: 8–20 mg/kg/day divided every 6–12 h.
    - Considerations: monitor creatinine and potassium in patients on high-dose therapy.
54. Vancomycin: Antibiotic, glycopeptide.
    - Systemic infection dosing:
      - Pediatric (IV):

    - 45–60 mg/kg/day divided every 6–8 h (monitor serum concentration)
  - Adult (IV):
    - 15–20 mg/kg/dose (rounded to nearest 250 mg) every 8–12 h (monitor serum concentration and/or trough levels)
- *C. difficile* dosing:
  - Pediatric (Oral):
    - 40 mg/kg/day divided every 6–8 h for 7–10 days (max 2 g/day)
  - Adult (Oral):
    - 125–500 mg 4 times daily
- Surgical prophylaxis: given 60 min prior to incision, repeat dose every 8 h.
  - Pediatric (IV):
    - Neonatal: 10 mg/kg (no repeat dosing).
    - Infants, children, adolescents: 15 mg/kg (max 1 g).
  - Adult (IV):
    - 15 mg/kg (max 2 g/dose)

55. Electrolyte *Repletion* (not to be used for maintenance):
    - Calcium:
      - Notes
        - Hypomagnesemia is a common cause of hypocalcemia, if serum magnesium concentration is low, this should be replaced prior to calcium depletion in most cases.
        - If IV calcium in required, calcium gluconate is typically preferred over calcium chloride because it is less likely to cause tissue necrosis if extravasated.
        - Milder cases of hypocalcemia may be treated with oral elemental calcium given as calcium carbonate or calcium citrate.
      - Calcium gluconate (Preferred):
        - Pediatric:
          - Oral: 30-75 mg/kg/day (elemental calcium) divided in 4–5 doses.
          - IV: 29–60 mg/kg/dose every 6 h.
          - Continuous IV: 8–13 mg/kg/h.
        - Adult (IV):
          - 1–4 g over 2–4 h (asymptomatic patients may be given oral)
      - Calcium carbonate (Tums):

    - Pediatric (Oral):
      - 45–65 mg/kg/day (elemental calcium) in 4 divided doses
    - Adult (Oral):
      - 500–4000 mg/day divided 1–3 times daily
  - Calcium citrate (Calcitrate OTC):
    - Pediatric (Oral):
      - 45–65 mg/kg/day (elemental calcium) in 4 divided doses
    - Adult (Oral):
      - 200–1000 mg/day (elemental calcium) divided 1–2 times daily
- Sodium:
  - Pediatric (IV):
    - Asymptomatic, mild, or moderate symptoms:
      - 0.9% isotonic solution over 24–48 h (max correction rate 10 mEq/24 h for acute hyponatremia)
    - Severe or neurologic symptoms:
      - 3% hypertonic solution bolus 2–5 mL/kg over 20 min (max 150 mL/dose), can repeat dose 2 times if severe symptoms continue (max correction rate 0.5 mEq/h or 10 mEq/24 h for acute hyponatremia)
  - Adult (IV):
    - Asymptomatic (serum sodium <130 mEq/L):
      - 3% hypertonic solution 50 mL bolus over 5+ min, monitor every 1–2 h to assess for additional treatment needs (max correction rate 8 mEq/24 h)
    - Symptomatic (serum sodium <130 mEq/L):
      - 3% hypertonic saline 100 mL bolus over 10 min, can repeat up to total of 3 doses over 30 min, monitor serum sodium hourly to assess for additional treatment needs (max correction rate 10 mEq/24 h)
- Magnesium:
  - Notes
    - Up to 50% of infused magnesium will be excreted in the urine due to inhibition of magnesium reabsorption in the loop of Henle caused by the infusion.

- If given IV, beware of respiratory depression, hypotension, heart block, or hypermagnesemia, calcium gluconate via IV may be used as antidote.
- Normomagnesemic magnesium depletion (cellular magnesium depletion) should be considered as a possible cause of refractory hypokalemia or unexplained hypocalcemia.

- Magnesium Sulfate:
  - Pediatric (IV):
    - 25–50 mg/kg/dose every 6 h for 2–3 doses, then recheck serum concentration (max 2 g/dose)
  - Adult:
    - Mild deficiency:
      1. IV: 1–2 g over 1–2 h.
      2. IM (50% concentration): 1 g every 6 h for 4 doses.
    - Moderate deficiency:
      1. IV: 2–4 g over 2–12 h.
      2. IM (50% concentration): 1 g every 6 h for 4 doses.
    - Severe deficiency:
      1. IV: 4–8 g over 4–24 h.

- Potassium:
  - Notes
    - IV potassium should be reserved for severe depletion, and continuous ECG monitoring should be utilized, monitor serum potassium closely.
    - Severe depletion or ongoing losses may require > 200% of normal daily limit needs.
    - Consider alternative solutions for atypical presentations: potassium bicarbonate for hypokalemia with metabolic acidosis or potassium phosphate for hypokalemia with hypophosphatemia.
  - Potassium chloride:
    - Pediatric:
      - Oral: 2–5 mEq/kg/day in divided doses (not to exceed 2 mEq/kg as single dose or 20 mEq, whichever is less).
      - IV: 0.5–1 mEq/kg/dose (infused at ≤0.5 mEq/kg/h, max 40 mEq/dose).
    - Adult:
      - Oral: 10–20 mEq 2–4 times daily (max 40 mEq/dose).

        - IV: 20 mEq every 2–3 h (max infusion rate 10–20 mEq/h.

- Phosphate:
    - Notes
        - Asymptomatic patients with serum phosphate less than 2 mg/dL may be treated with oral phosphate.
        - Serum phosphate levels below 1 mg/dL are typically treated with IV phosphate, switching to oral therapy when serum phosphate exceeds 1.5 mg/dL.
        - Consider halting repletion when serum phosphate is greater than or equal to 2 mg/dL.
        - Contact Children's pharmacy for available/appropriate IV dilution.
    - Potassium phosphate and sodium phosphate:
        - Pediatric (Oral):
            - 2–3 mmol/kg/day divided in 4+ doses
        - Adult (Oral):
            - 250–500 mg 4 times daily
    - Potassium phosphate:
        - Pediatric (IV):
            - Serum phosphorus 2.3–2.7 mg/dL:
                i. 0.08–0.16 mmol/kg over 4–6 h
            - Serum phosphorus 1.5–2.2 mg/dL:
                i. 0.16–0.32 mmol/kg over 4–6 h
            - Serum phosphorus <1.5 mg/dL:
                i. 0.32–0.64 mmol/kg over 4–6 h
        - Adult (IV):
            - Serum phosphorus 2.3–2.7 mg/dL:
                i. 0.08–0.16 mmol/kg over 4–6 h
            - Serum phosphorus 1.5–2.2 mg/dL:
                i. 0.16–0.32 mmol/kg over 4–6 h
            - Serum phosphorus <1.5 mg/dL:
                i. 0.32–0.64 mmol/kg over 4–6 h.

# Developmental Milestones

**Emily E. Steinhilber and Bryan S. Walters**

**Key Points**

1) There is a great variation in achievement in developmental milestones between individual children.
2) Loss of previous milestones is particularly worrisome and should be a cause to investigate the etiology.
3) Children should have regular visits with an established primary care provider so that progression through developmental milestones can be tracked as the child grows.

There are standardized charts for typical acquisition of developmental milestones in children. Clinicians must recognize that many factors can influence the age at which an individual child will attain milestones. Some factors are premature birth, nutrition, chronic illness, trauma, and recovery from operations or other treatments. The surgeon caring for a child will often be only seeing a snapshot in time for that child, so it is important to query the parents on the patient's developmental growth and usual behavior.

1. Up-to-date information on developmental milestones is available at www.cdc.gov/actearly or via 1-(800) CDC-INFO [1–3].

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2. Parents or providers can download the app “Milestone Tracker” from the CDC website at: https://www.cdc.gov/ncbddd/actearly/index.html [4].
3. Age: 2 months
    a. Motor: holds head up, smoother limb motion.
    b. Speech: coos, gurgles, turns to sound.
    c. Cognitive: attends faces, follows with eyes, gets bored with repetitive activity.
    d. Social: smiles, can briefly calm self, tries to look at parent.
4. Age: 4 months
    a. Motor: Holds head steady, pushes down with feet, rolls from prone to supine, shakes a toy, hands to mouth, when supine can prop on elbows.
    b. Speech: babbles, copies sounds, had different cries.
    c. Cognitive: expresses happy or sad, responds to affection, reaches out with one hand, directs hand to seen object, follows with eyes side to side, watches faces closely, recognizes people at distance.
    d. Social: smiles at people, likes to play, copies expressions.
5. Age: 6 months
    a. Motor: can roll supine to prone and back, sits unsupported, bounces on legs while standing, rocks back and forth on knees.
    b. Speech: responds to sound with sound, strings together vowels, responds to name, has happy and sad sounds, beginning consonants.
    c. Cognitive: looks around at things, brings things to mouth, is curious about objects, pass object from hand to hand.
    d. Social: can recognize a stranger as such, enjoys play, responds to emotions, gazes in mirror.
6. Age: 9 months
    a. Motor: stands, can sit up, can pull self to standing, crawls.
    b. Speech: understands “no,” “mama,” “baba” noises, copies sounds and gestures, points.
    c. Cognitive: watches falling objects, looks for hidden objects, peek-a-boo, puts things in mouth, smooth passing from hand to hand, pincer grasp.
    d. Social: fear of strangers, clings to know persons, has favorite toys.
7. Age: 1 year
    a. Motor: can sit up unaided, cruises on furniture, isolated unsupported steps, stands alone.
    b. Speech: responds to questions, can shake head no, can wave goodbye, can change tone of sounds, says “mama,” “dada,” tries to copy words.
    c. Cognitive: explores objects, finds hidden objects, knows names of pictures, uses simple objects as intended, bangs objects together, moves objects in and out of a container, releases objects, pokes, follows commands.

   d. Social: shy with strangers, cries when parent gone, has favorites, fearful, hands book to adult for story, repeats sounds for attention, assists with putting on clothing, plays "patty-cake."

8. Age: 18 months

   a. Motor: walks alone, climbs steps, pulls toys, helps remove clothing, drinks from cup, uses spoon.
   b. Speech: says single words, says "no" and shakes head, points at desired object.
   c. Cognitive: knows purpose of everyday items, points for attention, pretends to feed doll, points to body parts, scribbles, follow verbal commands without gestures.
   d. Social: enjoys handing off objects, has temper tantrum, plays pretend, clings to parent in new situation, points out objects of interest, explores alone near parent.

9. Age: 2 years

   a. Motor: stands on tiptoe, kicks ball, runs, climbs furniture, walks stairs, throws ball, draws lines and circles.
   b. Speech: knows names, short sentences, follows two-step instructions, repeats conversation, points in a book.
   c. Cognitive: sorts shapes and colors, completes sentences and rhymes, plays make-believe, builds towers, prefers one hand, names pictures.
   d. Social: copies others, excited with other children, shows independence, disobeys, plays next to other children.

10. Age: 3 years

    a. Motor: climbs well, runs easily, rides tricycle, walks stairs with one foot per step.
    b. Speech: follows three-step instructions, names familiar things, understands prepositions, can name self, age, and gender, names a friend, uses pronouns and plurals, can be understood by stranger, can converse.
    c. Cognitive: works moving parts, completes four-piece puzzle, understands "two," copies circle, turns page, builds six block tower, opens jar and door.
    d. Social: affection for friends, takes turns, has concern for crying friend, understands possession, has range of emotions, can separate from parent, upset with change in routine, can dress/undress.

11. Age: 4 years

    a. Motor: hops on one foot, catches bounced ball, pours, cuts, and mashes food.
    b. Speech: basic grammar and gender, sings song, recites poem, tells story, states first and last name.
    c. Cognitive: names colors and numbers, understands counting, understands time, remembers story, understands same/different, uses scissors, copies letters, plays board or card games, anticipates events in story.

   d. Social: enjoys new things, plays “mom” or “dad,” creative make-believe, prefers playing with others, cooperates with playmates, confuses real and make-believe, discusses likes and interests.

12. Age: 5 years
    a. Motor: Stands on one foot ten seconds, hops and skips, can do a somersault, uses fork and spoon, uses toilet, swings.
    b. Speech: speaks clearly, tells story with sentences, uses future tense, states name and address.
    c. Cognitive: counts to ten, draws person with six body parts, prints letters and numbers, copies triangle, understands money and food.
    d. Social: wants to be like and please friends, agrees with rules, sings, dances, acts, aware of gender, differentiates reality and make-believe, increased independence, can be demanding and cooperative at different times.

## References

1. Centers for Disease Control and Prevention, (CDC). http://www.cdc.gov/NCBDDD/ACTEARLY/milestones/ , Bright Futures: guidelines for health supervision of infants, children, and adolescents, 3rd ed. American Academy of Pediatrics, Elk Grove Village, IL, 2008.
2. Shelov S, Altmann TR. American academy of pediatrics. Caring for your baby and young child: birth to Age 5. 5th ed. London, UK: Bantam; 2009.
3. Hagan J Jr, Shaw JS, Duncan PM. American academy of pediatrics. Bright futures: guidelines for health supervision of infants, children, and adolescents, Third edition. American Academy of Pediatrics. Washington, DC: Elk Grove Village, IL, and US Department of Health and Human Services; 2008.
4. Moodie S, Daneri P, Goldhagen S, Halle T, Green K, LaMonte L. Watch me Thrive! Early childhood developmental screening: a compendium of measures for children ages birth to five. (OPRE Report 201411). Washington, DC: Office of Planning, Research and Evaluation, Administration, U.S. Department of Health and Human Services, www.cdc.gov/actearly. Accessed 3 NOV 2021.
5. Learn the signs. Act Early. Centers for Disease Control and Prevention, U.S. Department of Health and Human Services, Washington, DC. https://www.cdc.gov/ncbddd/actearly/index.html. Accessed 3 NOV 2021.

# Index Case List

**Christopher P. Coppola**

**Key Points**

1) The Accreditation Council for Graduate Medical Education (ACGME) sets case number minimums for general surgery residents with 20 pediatric surgery cases as part of the 850 total major cases.
2) The ACGME Pediatric Surgery Review Committee for Surgery sets a total major case requirement of 800 cases for graduating pediatric surgery fellows.
3) In a 2016 study, practicing pediatric surgeons reported that their most common cases were appendectomy, non-operative trauma care, and hernia repair while average numbers for 6 of 10 "rare" index cases were less than two per year for each surgeon [1].

1. ACGME case requirements for general surgery residents [2]
    a. Graduating residents must log a minimum 20 pediatric surgery cases, out of a minimum of 850 total major cases.
    b. Tracked pediatric surgery cases
        i. Bronchoscopy/esophagoscopy
        ii. Esophageal atresia/tracheoesophageal fistula
        iii. Gastroesophageal reflux (fundoplication)
        iv. Lung resection
        v. Congenital diaphragmatic hernia repair
        vi. Infantile hypertrophic pyloric stenosis

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        vii. Intestinal malrotation
        viii. Operative reduction of intussusception
        ix. Intestinal atresia, including duodenal atresia
        x. Appendectomy
        xi. Hirschsprung's disease (pull through)
        xii. Correction of anorectal malformation
        xiii. Operative correction biliary atresia
        xiv. Choledochal cyst
        xv. Gastroschisis/omphalocele
        xvi. Inguinal hernia repair <6 months
        xvii. Orchiopexy
        xviii. Ovarian and fallopian tube operations
        xix. Head and neck procedures
        xx. Non-operative management of multisystem trauma
        xxi. Neonatal cases (<44 weeks gestational age)
        xxii. Operative resection of tumors (including Wilms and neuroblastoma)

    c. A 2015 study of case numbers at a facility with general surgery residents and a pediatric surgery fellow demonstrated that the current list of tracked cases makes it difficult for all residents to meet the minimum case requirements. [3]

        i. The most common cases logged were pyloromyotomy, inguinal hernia, and umbilical hernia.
        ii. Inclusion of laparoscopic appendectomy and laparoscopic gastrostomy tube placement in the tracked cases would increase the chance that all residents would meet the requirement.

2. The ACGME Pediatric Surgery Review Committee for Surgery changed requirements for graduating pediatric surgery fellows starting in 2021[4].

    a. Case log requirements

        i. Total major case requirement: 800, with minimum numbers of complex cases
        ii. 50 teaching assistant cases

    b. Case category required minimums

        i. Abdomen/GI: 120
        ii. Thoracic: 50

            1. 30 additional endoscopies are required.
            2. 80 total procedures are required, but procedures performed on the same patient can be counted separately.

        iii. Trauma/critical care

            1. Operative trauma cases: 20
            2. Non-operative trauma management: 90

3. NICU managed cases: 20
4. PICU managed cases: 10
5. Oncology cases: 25
6. Head and neck/endocrine, genitourinary, and ano-recto malformation: 50

## Additional Notes

It remains an important part of general surgery residency training to include pediatric surgery cases. In all environments general surgeons will face questions on surgical issues of children, most notably after injury. In some practice settings, general surgeons will operate on children. A 2013 study showed a steady decline in the number of pediatric surgery cases performed by general surgery residents, notably associated with a decreasing number of hernia repairs performed. [5] Laparoscopic cases on children are counted as laparoscopy, not pediatric surgical cases, so one author has suggested including these cases as pediatric to better reflect the current emphasis on minimally invasive techniques in children [3]. After training, there is a challenge in maintaining competency in performance of index pediatric surgery cases. A review of case logs reported by recertifying pediatric surgeons demonstrated low numbers of cases per surgeon in categories of rare index cases. [1] The decreasing trend of number of cases per surgeon may be due to increased numbers of pediatric surgeons in practice, specialization of surgeons, and concentration of care in urban centers.

## References

1. Abdullah F, Salazar J, Gause C, et al. Understanding the operative experience of the practicing pediatric surgeon implications for training and maintaining competency. JAMA Surg. 2016;151(8):735–41.
2. Review Committee for Surgery, Defined category minimum numbers for general surgery residents and credit role, ACGME, 2019 (downloaded on 5/5/2021 from https://www.acgme.org/Portals/0/DefinedCategoryMinimumNumbersforGeneralSurgeryResidentsandCreditRole.pdf).
3. Thomas T, Reddy R, Ballard T, et al. ACGME case requirements for general surgeons in pediatric surgery: evaluation of current operative volume on resident completion of index cases. J Am Coll Surg. 2015;221(4, S1):S48.
4. Review Committee for Surgery, Defined category minimum numbers: pediatric surgery, ACGME, 2020 (downloaded on 5/5/2021 from https://www.acgme.org/Portals/0/PDFs/Pediatric_Surgery_Mapping.pdf).
5. Gow KW, Drake FT, Arabi S, Waldhausen JH. The ACGME case log: general surgery resident experience in pediatric surgery. J Peadiatr Surg. 2013;48(8):1643–9.

# Vaccination Schedule

**Emily E. Steinhilber and Bryan S. Walters**

1. Vaccination schedule for children and adolescents from the Centers for Disease Control and Prevention (CDC) [1] (Fig. 1).
2. The most recent vaccination information can be found at: http://www.cdc.gov/vaccines
3. Children who are not up to date on vaccinations should receive vaccinations on the catch-up immunization schedule[1] (Fig. 2).
4. Full footnotes available at: https://www.cdc.gov/vaccines/schedules/downloads/child/0-18yrs-child-combined-schedule.pdf
5. Vaccines, minimum age of administration, and recommended schedule.
    a. Hepatitis B (HepB)
        i. Minimum age: birth.
        ii. 3-dose series: Birth/1–2 months/6–18 months.
    b. Rotavirus (RV)
        i. Minimum age: 6 weeks.
        ii. 2-dose series: 2 months/4 months Rotarix (Rotavirus vaccine, live, oral. Glaxo Smith Kline, Philadelphia, USA).
        iii. 3-dose series: 2 months/4 months/6 months (RotaTeq).
    c. Diphtheria/tetanus toxoids/acellular pertussis (DtaP)
        i. Minimum age: 6 weeks.
        ii. 5-dose series: 2 months/4 months/6 months/15 months/4 years.

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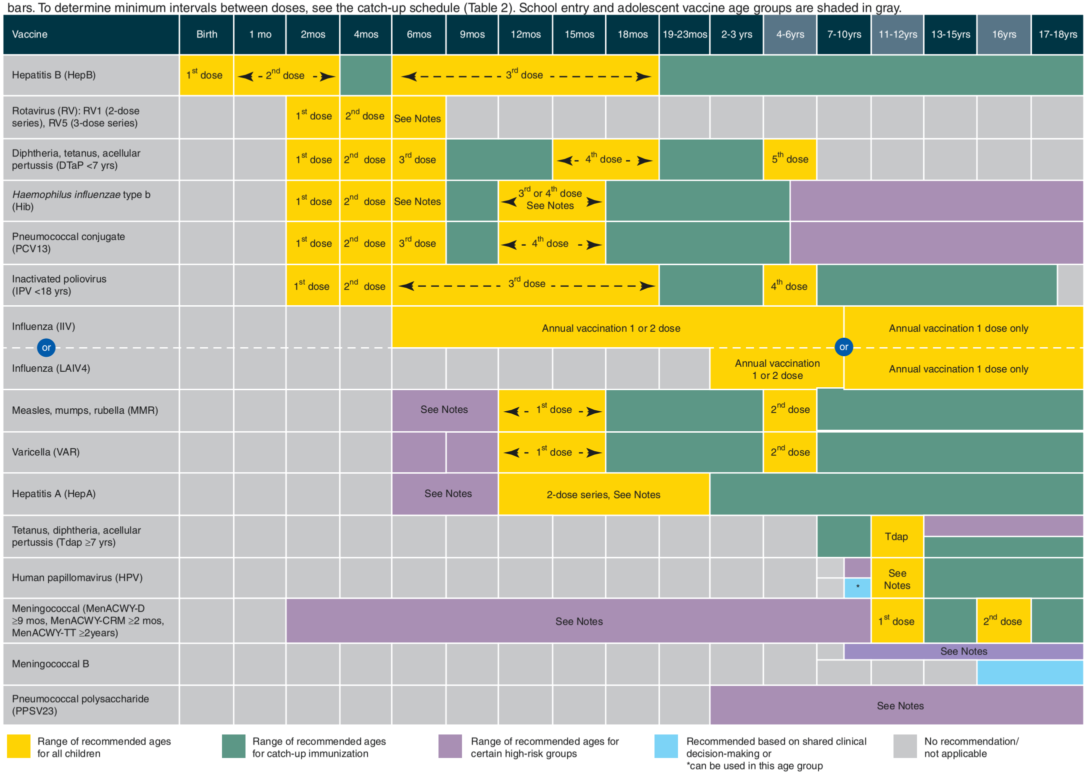


**These recommendations must be read with the notes that follow.** For those who fall behind or start late, provide catch-up vaccination at the earliest opportunity as indicated by the green bars. To determine minimum intervals between doses, see the catch-up schedule (Table 2). School entry and adolescent vaccine age groups are shaded in gray.

| Vaccine | Birth | 1 mo | 2mos | 4mos | 6mos | 9mos | 12mos | 15mos | 18mos | 19-23mos | 2-3 yrs | 4-6yrs | 7-10yrs | 11-12yrs | 13-15yrs | 16yrs | 17-18yrs |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Hepatitis B (HepB) | 1st dose | ← - 2nd dose - → | | 🟩 | ← - - - - - - - 3rd dose - - - - - - - → | | | | | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 |
| Rotavirus (RV): RV1 (2-dose series), RV5 (3-dose series) | ⬜ | ⬜ | 1st dose | 2nd dose | See Notes | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ |
| Diphtheria, tetanus, acellular pertussis (DTaP <7 yrs) | ⬜ | ⬜ | 1st dose | 2nd dose | 3rd dose | 🟩 | 🟩 | ← - 4th dose - → | | 🟩 | 🟩 | 5th dose | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ |
| *Haemophilus influenzae* type b (Hib) | ⬜ | ⬜ | 1st dose | 2nd dose | See Notes | 🟩 | ← 3rd or 4th dose See Notes → | | 🟩 | 🟩 | 🟩 | 🟩🟪 | 🟪 | 🟪 | 🟪 | 🟪 | 🟪 |
| Pneumococcal conjugate (PCV13) | ⬜ | ⬜ | 1st dose | 2nd dose | 3rd dose | 🟩 | ← - 4th dose - → | | 🟩 | 🟩 | 🟩 | 🟩🟪 | 🟪 | 🟪 | 🟪 | 🟪 | 🟪 |
| Inactivated poliovirus (IPV <18 yrs) | ⬜ | ⬜ | 1st dose | 2nd dose | ← - - - - - - - 3rd dose - - - - - - - → | | | | | 🟩 | 🟩 | 4th dose | 🟩 | 🟩 | 🟩 | 🟩 | 🟩⬜ |
| Influenza (IIV) | ⬜ | ⬜ | ⬜ | ⬜ | Annual vaccination 1 or 2 dose | | | | | | | | 🟨 Annual vaccination 1 dose only | | | | |
| or | | | | | | | | | | | | | or | | | | |
| Influenza (LAIV4) | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | Annual vaccination 1 or 2 dose | | 🟨 Annual vaccination 1 dose only | | | | |
| Measles, mumps, rubella (MMR) | ⬜ | ⬜ | ⬜ | ⬜ | See Notes | | ← - 1st dose - → | | 🟩 | 🟩 | 🟩 | 2nd dose | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 |
| Varicella (VAR) | ⬜ | ⬜ | ⬜ | ⬜ | 🟪 | 🟪 | ← - 1st dose - → | | 🟩 | 🟩 | 🟩 | 2nd dose | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 |
| Hepatitis A (HepA) | ⬜ | ⬜ | ⬜ | ⬜ | See Notes | | 2-dose series, See Notes | | | | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 | 🟩 |
| Tetanus, diphtheria, acellular pertussis (Tdap ≥7 yrs) | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | 🟩 | Tdap | 🟪🟩 | 🟪🟩 | 🟪🟩 |
| Human papillomavirus (HPV) | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜🟪<br>⬜🟦 * | See Notes | 🟩 | 🟩 | 🟩 |
| Meningococcal (MenACWY-D ≥9 mos, MenACWY-CRM ≥2 mos, MenACWY-TT ≥2years) | ⬜ | ⬜ | See Notes | | | | | | | | | | | 1st dose | 🟩 | 2nd dose | 🟩 |
| Meningococcal B | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜🟪<br>⬜ | See Notes<br>⬜ | 🟪<br>⬜ | 🟪<br>🟦 | 🟪<br>🟦 |
| Pneumococcal polysaccharide (PPSV23) | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | ⬜ | See Notes | | | | | | |

🟨 Range of recommended ages for all children

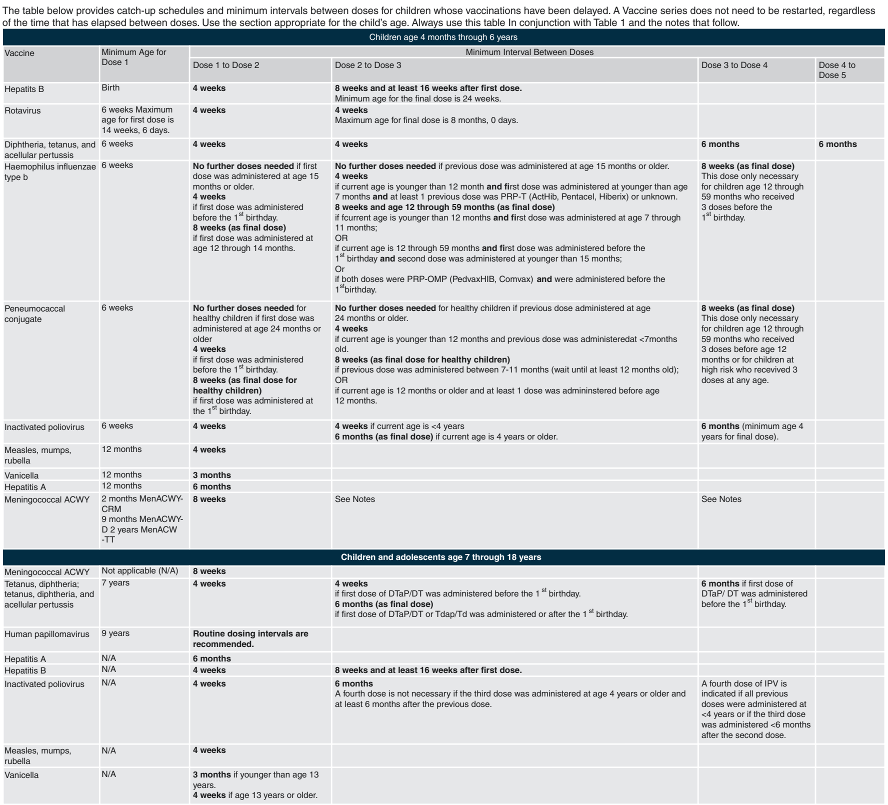
🟩 Range of recommended ages for catch-up immunization

🟪 Range of recommended ages for certain high-risk groups

🟦 Recommended based on shared clinical decision-making or<br>*can be used in this age group

⬜ No recommendation/ not applicable

**Fig. 1** Recommended Child and Adolescent Immunization Schedule for ages 18 or younger, United States, 2021. Source: Centers for Disease Control and Prevention, US Department of Health and Human Services. https://www.cdc.gov/vaccines/schedules/hcp/imz/child-adolescent.html, Accessed 21 OCT 2021. U.S. Government work in public domain



The table below provides catch-up schedules and minimum intervals between doses for children whose vaccinations have been delayed. A Vaccine series does not need to be restarted, regardless of the time that has elapsed between doses. Use the section appropriate for the child's age. Always use this table In conjunction with Table 1 and the notes that follow.

| Children age 4 months through 6 years | | | | | |
|---|---|---|---|---|---|
| Vaccine | Minimum Age for Dose 1 | Minimum Interval Between Doses | | | |
| | | Dose 1 to Dose 2 | Dose 2 to Dose 3 | Dose 3 to Dose 4 | Dose 4 to Dose 5 |
| Hepatits B | Birth | **4 weeks** | **8 weeks and at least 16 weeks after first dose.**<br>Minimum age for the final dose is 24 weeks. | | |
| Rotavirus | 6 weeks Maximum age for first dose is 14 weeks, 6 days. | **4 weeks** | **4 weeks**<br>Maximum age for final dose is 8 months, 0 days. | | |
| Diphtheria, tetanus, and acellular pertussis | 6 weeks | **4 weeks** | **4 weeks** | **6 months** | **6 months** |
| Haemophilus influenzae type b | 6 weeks | **No further doses needed** if first dose was administered at age 15 months or older.<br>**4 weeks**<br>if first dose was administered before the 1st birthday.<br>**8 weeks (as final dose)**<br>if first dose was administered at age 12 through 14 months. | **No further doses needed** if previous dose was administered at age 15 months or older.<br>**4 weeks**<br>if current age is younger than 12 month **and fi**rst dose was administered at younger than age 7 months **and** at least 1 previous dose was PRP-T (ActHib, Pentacel, Hiberix) or unknown.<br>**8 weeks and age 12 through 59 months (as final dose)**<br>if fcurrent age is younger than 12 months **and fi**rst dose was administered at age 7 through 11 months;<br>OR<br>if current age is 12 through 59 months **and fi**rst dose was administered before the 1st birthday **and** second dose was administered at younger than 15 months;<br>Or<br>if both doses were PRP-OMP (PedvaxHIB, Comvax) **and** were administered before the 1stbirthday. | **8 weeks (as final dose)**<br>This dose only necessary for children age 12 through 59 months who received 3 doses before the 1st birthday. | |
| Peneumocaccal conjugate | 6 weeks | **No further doses needed** for healthy children if first dose was administered at age 24 months or older<br>**4 weeks**<br>if first dose was administered before the 1st birthday.<br>**8 weeks (as final dose for healthy children)**<br>if first dose was administered at the 1st birthday. | **No further doses needed** for healthy children if previous dose administered at age 24 months or older.<br>**4 weeks**<br>if current age is younger than 12 months and previous dose was administeredat <7months old.<br>**8 weeks (as final dose for healthy children)**<br>if previous dose was administered between 7-11 months (wait until at least 12 months old);<br>OR<br>if current age is 12 months or older and at least 1 dose was admininstered before age 12 months. | **8 weeks (as final dose)**<br>This dose only necessary for children age 12 through 59 months who received 3 doses before age 12 months or for children at high risk who recevived 3 doses at any age. | |
| Inactivated poliovirus | 6 weeks | **4 weeks** | **4 weeks** if current age is <4 years<br>**6 months (as final dose)** if current age is 4 years or older. | **6 months** (minimum age 4 years for final dose). | |
| Measles, mumps, rubella | 12 months | **4 weeks** | | | |
| Vanicella | 12 months | **3 months** | | | |
| Hepatitis A | 12 months | **6 months** | | | |
| Meningococcal ACWY | 2 months MenACWY-CRM<br>9 months MenACWY-D 2 years MenACW -TT | **8 weeks** | See Notes | See Notes | |
| **Children and adolescents age 7 through 18 years** | | | | | |
| Meningococcal ACWY | Not applicable (N/A) | **8 weeks** | | | |
| Tetanus, diphtheria; tetanus, diphtheria, and acellular pertussis | 7 years | **4 weeks** | **4 weeks**<br>if first dose of DTaP/DT was administered before the 1 st birthday.<br>**6 months (as final dose)**<br>if first dose of DTaP/DT or Tdap/Td was administered or after the 1 st birthday. | **6 months** if first dose of DTaP/ DT was administered before the 1st birthday. | |
| Human papillomavirus | 9 years | **Routine dosing intervals are recommended.** | | | |
| Hepatitis A | N/A | **6 months** | | | |
| Hepatitis B | N/A | **4 weeks** | **8 weeks and at least 16 weeks after first dose.** | | |
| Inactivated poliovirus | N/A | **4 weeks** | **6 months**<br>A fourth dose is not necessary if the third dose was administered at age 4 years or older and at least 6 months after the previous dose. | A fourth dose of IPV is indicated if all previous doses were administered at <4 years or if the third dose was administered <6 months after the second dose. | |
| Measles, mumps, rubella | N/A | **4 weeks** | | | |
| Vanicella | N/A | **3 months** if younger than age 13 years.<br>**4 weeks** if age 13 years or older. | | | |

**Fig. 2** Recommended Catch-up Immunization Schedule for Children and Adolescents Who Start Late or Who Are More than 1-month Behind, United States, 2021. Source: Centers for Disease Control and Prevention, US Department of Health and Human Services. https://www.cdc.gov/vaccines/schedules/hcp/imz/child-adolescent.html, Accessed 21 OCT 2021. U.S. Government work in public domain

d. Haemophilus influenza type b (Hib)
   i. Minimum age: 6 weeks.
   ii. 4-dose series: 2 months/4 months/6 months/12–15 months.

e. Pneumococcal conjugate (PCV13)
   i. Minimum age: 6 weeks.
   ii. 4-dose series: 2 months/4 months/6 months/12–15 months.

f. Inactivated poliovirus (IPV)
   i. Minimum age: 6 weeks.
   ii. 4-dose series: 2 months/4 months/6–18 months/4–6 years.

g. Influenza (IIV)
    i. Minimum age: 6 months.
    ii. 6 months (annual, 1 or 2 doses).

h. Measles/mumps/rubella (MMR)
    i. Minimum age: 1 year.
    ii. 2-dose series: 12–15 months/4–6 years.

i. Varicella (VAR)
    i. Minimum age: 1 year.
    ii. 2-dose series: 12–15 months/4–6 years.

j. Hepatitis A (HepA)
    i. Minimum age: 12 months.
    ii. 2-dose series: 12 months/18 months.

k. Tetanus and diphtheria toxoids and acellular pertussis (Tdap)
    i. Minimum age: 11 years.
    ii. 1-dose series: 11–12 years.

l. Human papillomavirus (HPV)
    i. Minimum age: 9 years.
    ii. 11 years (2 or 3 dose series).

m. Meningococcal serogroup A, C, W, Y (MenACWY-CRM)
    i. Minimum age: 2 months.
    ii. 2 dose series at 11–12 years/16 years.

n. Meningococcal serogroup B (MenB-4C)
    i. Minimum age: 10 years.
    ii. Bexsero: 2-dose series at least 1 month apart: preferred age 16–18 years.
    iii. Trumenba: 2-dose series at least 6 months apart: preferred age 16–18 years.

o. Pneumococcal polysaccharide (PPSV23)
    i. Minimum age: 2 years.
    ii. 1-dose at least 8 weeks after completion of PCV13: 2–5 years.
    iii. After age 2 years for certain high-risk groups (asplenia, sickle cell disease, immunocompromise, transplant recipient, chronic heart disease, chronic lung disease, diabetes, cerebrospinal fluid leak, cochlear implant).

6. COVID vaccination
    a. The CDC recommends that everyone aged 12-years-old or older receives a COVID vaccine [2].

b. Vaccines approved by the Food and Drug Administration (FDA) in the USA are Pfizer-BioNTech, Moderna, Janssen/Johnson&Johnson.
    i. Pfizer-BioNTech
        1. Minimum age: 12 years. (On 27 OCT 2021, the FDA Vaccine advisory panel voted to allow this vaccine for ages 5-years-old and up.) [3].
        2. 2-dose series separated by 21 days.
        3. Moderately to severely immunocompromised people: Consider an additional dose at least 28 days after the initial 2-dose primary series.
    ii. Moderna
        1. Minimum age: 18 years.
        2. 2-dose series separated by 28 days.
        3. Moderately to severely immunocompromised people: Consider an additional dose at least 28 days after the initial 2-dose primary series.
    iii. Janssen/Johnson&Johnson
        1. Minimum age: 18 years and older.
        2. 1-dose series.

c. In select groups with increased risk, a booster shot is available [4].
    i. 65 years of age and older.
    ii. 18 through 64 years of age at high risk of severe COVID-19.
    iii. 18 through 64 years of age with frequent institutional or occupational exposure to SARS-CoV-2.

d. Government regulations require that COVID-19 vaccination be provided without any cost to recipients and may not be denied based on insurance coverage.

e. Recommendations are rapidly evolving. The latest recommendations from the CDC can be found at: https://www.cdc.gov/coronavirus/2019-ncov/vaccines/keythingstoknow.html

## References

1. Recommended Child and Adolescent Immunization Schedule for ages 18 years or younger, United States, 2021 Centers for Disease Control and Prevention, US Department of Health and Human Services. https://www.cdc.gov/vaccines/schedules/hcp/imz/child-adolescent.html. Accessed 21 OCT 2021. U.S. Government work in public domain.
2. Key Things to know about COVID-19 vaccines. Centers for Disease Control and Prevention, US Department of Health and Human Services. https://www.cdc.gov/coronavirus/2019-ncov/vaccines/keythingstoknow.html. Accessed 21 OCT 2021. U.S. Government work in public domain.

3. Fox M, FDA vaccine advisors vote to recommend Pfizer's Covid-19 vaccine for children 5 to 11. CNN health. Cable News Network, 2021. https://www.cnn.com/2021/10/26/health/covid-19-young-kids-vaccine-fda-discussion/index.html. Accessed 27 OCT 2021.
4. Coronavirus (COVID-19) Update: FDA Takes Additional Actions on the Use of a Booster Dose for COVID-19 Vaccines. https://www.fda.gov/news-events/press-announcements/coronavirus-covid-19-update-fda-takes-additional-actions-use-booster-dose-covid-19-vaccines. Accessed 21 OCT 2021. U.S. Government work in public domain.

# Phone List

| Resource | Phone | Contact | email |
|---|---|---|---|
| Admissions | | | |
| Adolescent Medicine | | | |
| Anesthesiology | | | |
| Blood Bank | | | |
| Button Battery Hotline | 800-498-8666 | | |
| Cafeteria | | | |
| Cardiology | | | |
| Cath Lab | | | |
| Chemistry | | | |
| Clinic | | | |
| CT | | | |
| Cytology | | | |
| Dermatology | | | |
| Disaster Distress Hotline | 800-985-5990 | | |
| Domestic Violence Hotline | 800-799-7233 | | |
| Eating Disorders Helpline | 800-931-2237 | | |
| ENT | | | |
| ER | | | |
| ER | | | |
| Frozen Section | | | |
| Gastroenterology | | | |
| Genetics | | | |
| Hematology | | | |
| Hematology | | | |
| Hemodialysis | | | |
| Human Trafficking Hotline | 888-373-7888 | | |
| Infectious Disease | | | |
| Interventional Radiology | | | |

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| | | | |
|---|---|---|---|
| Lab | | | |
| LGBT Hotline | 888-834-4564 | | |
| Medical Records | | | |
| Mental Health Help Line | 800-950-6264 | | |
| Microbiology | | | |
| MRI | | | |
| Narcotics Anonymous | 800-866-1463 | | |
| Neonatology | | | |
| Nephrology | | | |
| Neurology | | | |
| Neurosurgery | | | |
| NICU | | | |
| Nuclear Medicine | | | |
| Nutrition | | | |
| OB/GYN | | | |
| Occupational Therapy | | | |
| Oncology | | | |
| Operator | | | |
| Ophthalmology | | | |
| OR | | | |
| Orthopedics | | | |
| Paging Operator | | | |
| National Parent Helpline | 855-427-2736 | | |
| Pain Service | | | |
| Pathology | | | |
| Pediatric Surgery | | | |
| Pediatrics | | | |
| Perioperative Care | | | |
| Pharmacy | | | |
| Physical Therapy | | | |
| PICU | | | |
| Plastic Surgery | | | |
| Police | | | |
| Poison Control | 800-222-1222 | | |
| Psychiatry | | | |
| Pulmonary Lab | | | |
| Pulmonology | | | |
| Radiation Therapy | | | |
| Radiology | | | |
| Reading Room | | | |
| Rehabilitation | | | |
| Respiratory Therapy | | | |
| Runaway Hotline | 800-786-2929 | | |
| Sexual Assault Hotline | 800-656-4673 | | |
| Scheduling | | | |

| | | | |
|---|---|---|---|
| Security | | | |
| Social Work | | | |
| Speech Pathology | | | |
| Substance Abuse Hotline | 800-622-4357 | | |
| Suicide Prevention | 800-273-8255 | | |
| Surgical Pathology | | | |
| Thoracic Surgery | | | |
| Trans Lifeline | 877-565-8860 | | |
| Transplantation | | | |
| U/S | | | |
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| Vascular Lab | | | |
| X-ray File Room | | | |

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